Timing of Mask Fitting as a Predictor of Adherence in Children Requiring Non-Invasive Ventilation Therapy

By

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A thesis submitted in partial fulfillment of the requirements for the degree of

Master of Nursing

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Abstract

Non-invasive ventilation (NIV) therapy is an integral treatment for managing various pediatric disorders that disrupt breathing. Home NIV therapy has become increasingly prevalent in pediatric medicine, providing necessary chronic ventilation support and airway patency to prevent suboptimal ventilation. Appropriate mask fitting and headgear adaptation have been documented to be essential parts of the care and support for children requiring NIV; however, the effect on the timing of the mask fitting is not well studied in the literature. This thesis addresses this gap, investigating the impact of early mask fitting and headgear adaptation prior to NIV initiation and the potential benefits of maintaining adherence within six months post-NIV initiation. The first chapter provides background information on pediatric NIV. The second chapter presents the manuscript detailing the dissertation research project prepared for publication post-defense. The final chapter outlines the implications for future research and the practical implications for nursing. This research utilized secondary data analysis of retrospectively collected data from the pediatric NIV program at Stollery Children's Hospital between 2012 and 2015. The study did not find a significant impact on the timing of mask fitting for NIV initiation, adherence, or discontinuation at six months after NIV was initiated. Notably, adherence rates in both cases and controls were higher than previously reported in the literature, suggesting that mask fitting and headgear adaptation are key interventions for successful NIV initiation when performed before or shortly after therapy begins.

Keywords: Non-Invasive Ventilation Therapy, Mask Fitting, Adherence Rate, Pediatrics Respirology

Preface

This thesis is an original work by Ling (Lily) Lu. No part of this thesis has been previously published. 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 "Timing of mask fitting as a predictor of increased adherence in children requiring NIV Therapy," Pro00120218, August 4, 2022.

The data analyzed in this thesis was used with the permission of and is owned by Alberta Health Services (AHS). Study data were managed using Research Electronic Data Capture (REDCap), an electronic data capture tool 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 standard statistical packages; and 4) procedures for data integration and interoperability with external sources (1, 2).

Chapter 2 of this thesis will be ready for submission for publication. I was responsible for the data cleaning, analysis, and manuscript composition. Dr. Maria Castro Codesal and Dr. Shannon Scott co-designed the study; Deborah Olmstead, the lead of the pediatric NIV program at the Stollery Children's Hospital, provided the primary database from NIV clinic visits and from sleep company providers Sleep Medix and Medigas. The NIV adherence data was provided through their patient databases (3, 4). Dr. Castro Codesal and Dr. Scott were the supervisory authors and were involved with concept formation and manuscript composition.

Acknowledgments

I would like to express my heartfelt gratitude to my supervisors, Dr. Shannon Scott and Dr. Maria Castro Codesal, for their unwavering support and encouragement throughout my three years of research. Their passion for pediatric research was inspiring, and I feel privileged to have the opportunity to work with them.

I would also like to extend my appreciation to my thesis committee member Deborah Olmstead, Nurse Practitioner, for sharing her invaluable knowledge and expertise in pediatric NIV therapy. I am grateful for the opportunity to learn from her.

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

Lastly, I sincerely thank my husband, Bill Chan, my son, Gavin Chan, and my family for their constant support and encouragement throughout my graduate school journey. Their encouragement and belief in my abilities have been invaluable.

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Chapter 1: Introduction

Background on Non-Invasive Ventilation

Non-invasive ventilation (NIV) therapy is a mode of breathing support through a mask interface outside of the airway that is necessary for managing various pediatric disorders that disrupt breathing during sleep (5-7). Home NIV therapy offers pediatric patients an improved quality of life and well-being by providing necessary chronic ventilation support, maintaining airway patency, and preventing suboptimal ventilation during sleep (8-10). It also prevents the need for more invasive forms of ventilation and, ultimately, reduces respiratory morbidity and death (6, 7, 9, 11). Some indications for NIV include chronic respiratory failure, musculoskeletal weakness or chest wall restriction, obstructive sleep apnea, craniofacial malformations, airway malacias, sleep disorders associated with neurological conditions, and abnormalities in central respiratory drive (6-8, 10, 12-15). Appendix A includes definitions and explanations of NIV terminology used throughout the thesis.

The pediatric population requires focused research on NIV therapy, as children are unique physiologically (16). For example, infants' and children's airways and lungs are still developing, and their respiratory physiology highly depends on age, weight, and developmental stage (12). These factors add additional challenges related to NIV therapy and adherence, which include finding mask interfaces for pediatric face sizes and their growing faces, avoiding mask-related side effects, as well as tolerance issues to mask interfaces and air pressures (6, 11-13, 17). This requires healthcare practitioners to have an advanced understanding of the support for optimizing adherence in pediatric NIV therapy so that patients and families can receive evidence-informed, personalized care that improves NIV use and overall health for children.

Literature focused on NIV adherence rates in pediatric populations is disparate, with results suggesting factors associated with poor adherence, including poor mask fit, are partly related to limited options for pediatric interfaces and headgear, as well as the development of short and long-term complications associated with the therapy (7, 9, 12, 18-20). Complications from mask interfaces can jeopardize therapy use, either related to an inappropriate fit (e.g., skin injury, leaks, mucosal drying or excessive skin hydration, conjunctivitis, corneal ulcers) or the pressure exerted by the interface (e.g., skin erythema or ulcer, facial deformity, maxillary retrusion) (6, 17). However, many other factors (e.g., sex, race, age, BMI, etc.) may impact adherence rates (18, 21). For instance, there is a trend toward better NIV adherence with female sex, Caucasian race, younger ages, lower BMI, and having a mother with higher levels of education (18, 21). A higher baseline apnea-hypopnea index and a presence of developmental delay also tended to be associated with better adherence (18).

Appropriate mask and headgear fitting are crucial considerations to effectively initiate NIV and facilitate adherence while decreasing the likelihood of complications, as highlighted in the European Respiratory Society (ERS) guideline for NIV initiation in children published in 2022 and the 2018 Canadian Pediatric Home Mechanical Ventilation guideline (6, 9, 11, 15, 22, 23). However, to date, original research evaluating strategies to improve mask and headgear fit and, ultimately, improved adherence remains limited. This gap highlights the need to research whether the interventions to improve mask fit and the timing of these interventions can potentially shape outcomes for pediatric patients using NIV therapy.

NIV Initiation and Mask Fitting

Several standard mask interface types are available for home NIV use in the pediatric population (11, 14, 24). Figure 1 visually depicts the available types of mask interfaces (11).

Nasal masks are the most commonly used and preferred interfaces in the pediatric population for avoiding the risks of aspiration with emesis or asphyxia, lessening the likelihood of air leaks, and demonstrating less airway resistance (6, 24). Other interfaces, such as oronasal masks, nasal pillows, and total face masks, may be indicated in poor tolerance or side effects (e.g., mouth leak) from nasal masks or up to the patient's preference (11, 24). Although only used for diurnal NIV while awake, mouthpieces offer an alternative that prevents nasal symptoms and skin injuries (11, 24). When selecting a mask, it is important to consider the individual patient's needs and their risks for potential complications (6, 11, 24). Proper mask fitting is key to minimizing complications and ensuring effective therapy (6, 11, 24).

Figure 1. Types of Non-Invasive Ventilation Masks Available to Pediatric Patients in the Early 2010s.

Reproduced from Castro Codesal et al [11].

The pediatric population using home NIV therapy faces unique challenges due to the need for adaptation as they grow and the limited availability of commercially available mask options, particularly for infants (6, 11, 17, 24, 25). Also, specific subpopulations, such as children with syndromic craniofacial malformations require customized masks that accommodate their unique facial features (11, 24, 26-30). Current initiatives include exploring biocompatible soft materials and three-dimensional (3D) printing techniques to create personalized masks that minimize air leaks and improve comfort (11, 24, 26-30). Research in this area is growing, with studies showing reduced air leaks and improved comfort with 3D-

printed masks compared to conventional ones (26-30). However, these resources are still in the early stages of development, are costly and require further exploration and clinical testing before widespread implementation can be considered (26-30).

Headgear adaptation remains a critical aspect of ensuring a proper mask fit and stability, which is a component of mask fitting (6, 11, 24, 31). Appropriately fitted headgear stabilizes the mask interface and is essential for all children, and particularly in infants or children with skull or cranial deformities (11). The literature emphasizes the importance of using well-fitting headgear to avoid excessive tightening that can lead to skin injuries. Optimization of the headgear also stabilizes the NIV mask, preventing mask displacement and friction injuries (11, 24, 31). Innovative approaches, such as using different materials for headgear and developing customizable options, are being explored to improve comfort and effectiveness (11, 24). The "two-finger rule" for headgear tightness, along with regular assessment and adaptation of the headgear as the child grows, are recommended practices to minimize complications and enhance therapy adherence (11, 31). Research and clinical practices suggest continuous monitoring and early interventions to adjust the headgear can improve patient comfort and therapy outcomes (6, 11, 24, 31).

The lack of a properly fitting mask leads to side effects and ultimately to diminished tolerance to the NIV therapy and reduced adherence rates (6, 11, 14). Customization becomes imperative to ensure a proper fit and mitigate unwanted side effects for patients. Though there are documented benefits of these initiatives—such as improved adherence to therapy and a decrease in skin injuries among children (17, 25, 27), there remains a lack of research exploring the customization of headgear and the optimal timing for both mask fitting and headgear

adaptation. This gap persists despite the recognized potential impact on adherence rates and the development of complications, as indicated by expert opinion (6).

Adherence to NIV Therapy

Parameters defining adequate NIV adherence for the pediatric population lack universal validation and are often extrapolated from adult standards (6, 25). In adults, acceptable overnight NIV adherence typically involves usage for at least four hours a night for a minimum of five nights (70%) per week, a criterion frequently employed by public and private agencies for funding considerations (8, 14, 25). While this definition is commonly applied in pediatric literature, its limitations are acknowledged, particularly in younger children (<14 years) who may have longer sleep durations, aligning with recommendations from the American Academy of Sleep Medicine (32). The primary goal is optimal adherence, so using NIV throughout the entire sleep duration is advocated (6).

Currently, in North America, advanced technology allows pediatric patients and their families to monitor and upload their NIV adherence data wirelessly from their NIV machine to a cellphone application or via a secure digital memory (SD) card through a computer, making this data accessible to clinicians (33). Clinicians can then access the patients' adherence data through cloud-based clinician portals, where compliance reports, including adherence data, can be accessed online at any time (34). This technology enables clinicians to manage patients safely and effectively, facilitating remote monitoring of both short-term and long-term adherence while helping identify specific issues that may limit patients' tolerance (25, 34). However, during the study period between 2012 and 2015, a cloud-based system was not yet available for obtaining NIV adherence data. Instead, data was stored on SD cards and memory sticks that required patients and families to bring these to their sleep vendors for downloading prior to attending NIV

clinic visits (35). This data was then stored on the sleep companies' systems and faxed to the clinician's office.

Most adherence reports provide a 30-day report. Four adherence variables in the reports provide key metrics to assess different aspects of a child's NIV use and adherence, offering clinicians a comprehensive understanding of the consistency and effectiveness of therapy. **Average usage on total days** provides an overview of overall adherence by calculating the average duration of therapy used across all days in the observed period. This variable helps clinicians understand the consistency and frequency of therapy use over time. **Average usage on days used only** offers a more accurate reflection of how long the therapy is used on days it is utilized, excluding days with no usage. **Days used** is a count of the days the therapy was used during the observed period, indicating the frequency of usage. **Percent of days used over 4 hours** identifies the percentage of days during the observed period when the therapy was used for more than 4 hours. Extended usage is essential for achieving the full therapeutic benefits of NIV, making this metric a commonly used indicator by public and private agencies for funding considerations.

To illustrate the importance of reviewing all the adherence variables, consider two patients with adherence reports for 30 days. Patient A used NIV daily and had a compliance report showing an average use of 3 hours per day, an average use of 3 hours on days used only, a total of 30 days used, and 0% of days used over 4 hours. In contrast, Patient B used the therapy only on 10 days. His report also showed an average use of 3 hours per day, but an average use of 9 hours on days used, a total of 10 days used, and 33% of days used over 4 hours. In this example, Patient A used NIV consistently but perhaps faced some challenges with NIV adherence, whereas Patient B used NIV less often but for longer durations on the days it was

used. By examining all four variables, healthcare professionals can have a better understanding of different adherence patterns, investigate reasons for non-adherence, and formulate individualized strategies to optimize NIV adherence (35).

NIV adherence reports also contain detailed therapy information beyond adherence. For instance, bilevel positive airway pressure (BIPAP) reports provide information about direct measurements such as patient-triggered breath rate, inspiratory time, and reached pressures, as well as algorithm-calculated metrics such as tidal volume, leak, and the apnea-hypopnea index (25, 36). Although the accuracy of these parameters is limited, particularly in younger children with weight under the manufacturer's recommendations, these measures are useful for adjusting therapy settings and may ultimately impact therapy effectiveness and overall adherence.

While a standardized definition for NIV adherence rates in the pediatric population is lacking, and different adherence parameters might need to be used, earlier research emphasizes the predictive value of early adherence patterns for long-term outcomes (7, 18, 19, 37). Consequently, identifying and understanding early adherence patterns can provide valuable insights for healthcare practitioners caring for children requiring long-term NIV.

Complications to NIV Therapy

Improper mask fitting is one of the leading barriers to NIV adherence and a significant cause of complications among infants and children requiring NIV therapy (11, 14, 16, 24, 38, 39). Interface-related complications are common and can be categorized into several types: mask-related skin issues, short-term pressure-related injuries, humidification-related issues, leak-related issues, and long-term complications.

Mask-related skin issues can sometimes relate to prolonged use of NIV masks and can include persistent redness or more severe skin breakdown and pressure ulcerations (11, 14, 16,

24, 38, 39). Factors contributing to skin injury include excessive mask tightening, which creates pressure and friction on the face. Poor humidity can cause nasal congestion that makes NIV air pressures uncomfortable and poorly tolerated (11, 14, 16, 38, 39). Ensuring an optimal mask fit and providing small breaks (mask releases) when prolonged mask use is required are essential for preventing skin breakdown. Early detection of mask-related skin concerns and early preventive intervention, along with ongoing mask and headgear fitting as the child grows, are key to minimizing skin-related trauma (11, 14, 16, 24, 38, 39).

Short-term air pressure-related issues can include gastrointestinal bloating, and aspiration (12, 17, 18, 24). High airway pressures can cause gastric distention as air enters the stomach, which may lead to abdominal discomfort and increase the risk of aspiration. (16, 24, 39). Precautions such as using nasogastric tubes can help manage these risks (16, 24, 39).

Humidification-related issues can include nosebleeds, airway dryness leading to inflammation, and nasal congestion. These issues often arise from poor humidity or improper mask fitting. Nosebleeds and airway dryness can be reduced by using the lowest effective pressure and utilizing heated humidity (11, 14, 16, 38, 39).

Air leaks from the NIV interface can cause leak-related issues such as eye irritation and mouth leaks. Eye irritation is caused by air leaks reaching the eyes, while mouth leaks can disrupt effective therapy. These problems can be addressed by ensuring a proper mask fit and adapting headgear to reduce leaks (11, 14, 16, 24, 38, 39). Increased humidity and postural changes may also help manage air leaks (16, 17).

Long-term use of NIV can cause side effects such as pneumothorax and midface hypoplasia (11, 14, 16, 24, 38, 39). Although rare, pneumothorax is a serious complication caused by high inspiratory pressure, which can lead to lung over-distention (16, 17). To

minimize this risk, clinicians should monitor volume delivery and adjust settings for the lowest effective pressure (16, 17). Midface hypoplasia can occur in children using NIV long-term, usually caused by heavy masks or mask tightness with prolonged NIV use, particularly those with neuromuscular disorders (17, 24). Ongoing adjustments to mask fitting and headgear, along with regular evaluations of the maxilla-mandibular growth and early referral to pediatric orthodontists and maxillo-facial specialists, are crucial for preventing these deformities (11, 14, 16, 38, 39).

While NIV therapy is essential for many pediatric patients, it is accompanied by various complications that require careful management. Many complications, especially mask-related skin issues, may be minimized and even eliminated with optimal, customized equipment (11, 14, 16, 25, 38, 39). To our knowledge, no previous research has captured the impact of mask fitting and headgear customization and when it should be done to minimize complications.

Stollery's Pediatric NIV Program

Pediatric home NIV therapy is a highly specialized field. The Stollery Children's Hospital is a tertiary pediatric medical center with 236 beds located in Alberta, Canada, with a unique, interdisciplinary, and comprehensive pediatric home NIV program (40). The program is led by a nurse practitioner and joined by a team of pediatric respirologists and a respiratory therapist. It is also supported by social workers, occupational therapists (inpatient and outpatient), child life specialists and orthotists.

A vital aspect of the program is determining the need for individualized mask fitting and headgear customization prior to the recommended NIV therapy being initiated. The Stollery Children's Hospital's mask fit program was established through a recognized need to optimize NIV masks for infants and children when commercial headgear was not able to provide access to

appropriate pediatric mask interfaces. Initially, not every patient required mask fitting and headgear adaptation prior to initiating NIV therapy. NIV masks and instructions were provided by sleep companies, with patients and families expected to initiate therapy on their own. However, over the study period, there was a shift towards a more proactive approach of integrating mask fitting and headgear adaptation (if required) using available pediatric interfaces in infants and children prior to initiating NIV. This novel practice was established in order to optimize the mask fit, which in turn was anticipated to enhance the tolerance to and effectiveness of the therapy and reduce adverse effects from the outset.

Notably, the mask fit and headgear adaptation program at the Stollery Children's Hospital is unique and originated from an established need when standard headgear did not fit an infant or child referred to the NIV program. As noted, headgear is a key factor in mask stabilization, ensuring not only comfort but also usability for the child. During this process, occupational therapists and occupational therapy assistants thoroughly evaluate the patient's facial structure and NIV mask fit for a precise fit through customization of the headgear. This can include the adjustment of interface straps, the addition of non-skid materials to stabilize the headgear, and even utilizing several headgear types together in order to create a new modified headgear that optimizes the mask fit and relieves pressure points. Furthermore, custom helmets designed by orthotists are available for highly specialized patients, such as those with plagiocephaly. These helmets accommodate unique cranial shapes, enhancing mask fitting and ensuring more effective and comfortable NIV therapy, which would not be possible with regular headgear adaptations. Appendix B provides visual examples of good and poor NIV mask fit and headgear adaptation in both infant and child mannequins, illustrating the importance of proper sizing and fitting for effective NIV therapy.

This shift towards a proactive mask fitting and headgear adaptation at the Stollery Children's Hospital highlights the program's individualized patient care model and commitment to enhancing NIV adherence rates through optimization of the NIV mask fit. Over time, the team has identified that customized headgear optimizes mask fits and has become an essential component in facilitating increased NIV tolerance and use with minimal side effects. Currently, no literature can be found on the optimal timing for such interventions. The Stollery Children's Hospital's NIV team theorizes that commencing mask fitting early, prior to the initiation of home NIV, could positively affect initial and short-term adherence rates.

Purpose Statement and Research Questions

Purpose Statement

This study aims to evaluate whether timely interventions, specifically specialized mask fittings and headgear adaptations (if needed), are more effective in facilitating the initiation of NIV, enhancing adherence rates, and reducing discontinuation rates among an identified pediatric population using long-term home NIV.

Research Questions

The study is guided by three research questions:

1. Does early mask fitting and headgear adaptation (if needed) facilitate the initiation of NIV within the first six months following NIV recommendation?
2. Does early mask fitting and headgear adaptation (if needed) improve adherence to NIV at 1, 3, and 6 months after NIV initiation?
3. Does early mask fitting and headgear adaptation (if needed) impact NIV discontinuation within 6 months after NIV initiation?

Hypothesis

If pediatric patients from the Stollery Children's Hospital's NIV program receive early mask fitting and headgear adaptation (if needed) prior to NIV initiation, they are more likely to be successful in initiating NIV therapy and maintain adherence with less discontinuation to therapy at 1, 3, and 6 months after initiation.

Significance of the Study

In 2022, the ERS published one of the first statements by a professional respiratory society on pediatric long-term NIV (6). The statement highlighted that appropriate mask and headgear fitting are critical for initiating and maintaining NIV, and it indicated a need for more research on the importance of headgear and the use of complementary technologies to guide mask fitting (6). Additionally, the 2018 Canadian Pediatric Home Mechanical Ventilation guideline stated that for NIV, including mask ventilation or mouthpiece ventilation, interfaces need to be trialed to ensure comfort and effectiveness, which emphasizes the importance of mask fitting and headgear adaptation in NIV therapy (15).

Results from our study could potentially inform future research surrounding NIV therapy through the unique lens of mask fitting. The findings from this case-control research will examine the importance of early mask/headgear customization prior to NIV initiation as a key facilitator for adherence to therapy. Examining and describing the timing of mask fitting and headgear adaptation in the pediatric population could inform healthcare professionals to provide better care and potentially improve patient outcomes. This data can then help inform resource allocation and future program planning, ensuring that the pediatric NIV population is adequately supported. In addition, this study's findings could serve as a foundation for future home NIV therapy practices and help standardize the process involved in initiating pediatric patients into NIV therapy for optimal adherence. This foundational knowledge could also provide a

background for further research surrounding pediatric NIV therapy adherence and associated reduction in complications from NIV therapy.

Manuscript Overview

The following manuscript details a secondary analysis of a retrospective case-control sampled data provided by the pediatric NIV program at the Stollery Children's Hospital and the two sleep vendors, Sleep Medix and Medigas, between 2012 and 2015 (3, 4). Data cleaning was completed, and data analysis was done using SPSS. The manuscript will be submitted for publication after the thesis defense.

Chapter 2: Manuscript: Timing of Mask Fitting as a Predictor of Adherence in Children Requiring Non-Invasive Ventilation Therapy

Abstract

Introduction: The utilization of non-invasive ventilation (NIV) in the pediatric population is becoming increasingly prevalent. NIV is a form of breathing support that employs a mask interface outside the airway. It provides necessary respiratory support that improves patients' quality of life by maintaining airway patency and preventing inadequate ventilation during sleep. While appropriate mask fitting and headgear adaptation enhance NIV adherence, the impact of timing on these interventions is underexplored. This study examined the timing of mask fitting and headgear adaptation on NIV initiation, adherence, and discontinuation within six months post-initiation.

Methods: This retrospective, case-control study used secondary data analysis to examine pediatric patients aged 0-17 in the Stollery Children's Hospital NIV program who received mask fitting and headgear adaptation (if needed) between 2012 and 2015. Participants were divided into case and control groups based on the timing of mask fitting and headgear adaptation, either prior to initiating NIV (cases) or following initiation (controls). Demographic and clinical data were collected from the NIV program's clinical database, and adherence data were downloaded from patients' machines into a database provided by the sleep vendors. Outcomes of interest included NIV initiation rates, NIV usage at 1, 3, and 6 months, as well as NIV discontinuation rates at six months.

Results: One hundred and ten patients (29% female) were included, resulting in 146 mask-fitting entries (69% cases, 31% controls). The median NIV initiation age was 8 years, and upper airway conditions were the most common primary indication (64%). NIV was predominantly initiated in

outpatient settings (72%), with CPAP (63%) and nasal masks (91%) being most commonly used. 76% of patients initiated within six months after NIV was recommended based on clinical documentation. Based on available adherence reports, at one month, 54% of cases and 44% of controls initiated NIV (p = 0.341; OR: 1.48, 95% CI: 0.66–3.35), increasing to 57% and 50% by three months (p = 0.522; OR: 1.30, 95% CI: 0.58–2.93), and 61% and 53% by six months (p = 0.456; OR: 1.36, 95% CI: 0.60–3.08). When examining NIV usage over a 30-day period, at one month, cases used NIV for 4.6 hours per night (vs. 4.1 hours in controls; p=0.4), 5.7 hours (vs. 4.3 hours; p=0.13) at three months, and 6.3 hours (vs. 5.1 hours; p=0.51) at six months. The percentage of days with NIV use >4 hours was 56% in cases (vs. 45% in controls; p=0.29) at one month, 60% (vs. 46%; p=0.22) at three months, and 88% (vs. 61%; p=0.18) at six months. Dropout rates within the first 6 months were 15% in the case group and 22% in the control group, with no significant difference (p = 0.504; OR: 0.63, 95% CI: 0.16–2.48).

Conclusion: This study did not reveal a significant impact with respect to the timing of mask fitting on NIV initiation and therapy adherence at six months. This is not surprising since many other known factors can impact NIV adherence, including patient comfort, severity of medical conditions, and family involvement. This study, however, showed adherence rates in both cases and controls higher than previously reported in the literature, suggesting mask fitting and headgear adaptation are key interventions in successful NIV initiation, either before or shortly after NIV initiation. Since these interventions have become the standard of care, collecting further information on the control group for further comparisons will be challenging.

Keywords: Non-Invasive Ventilation Therapy, Mask Fitting, Adherence Rate, Pediatrics Respirology

Background on Non-Invasive Ventilation

Non-invasive ventilation (NIV) therapy provides breathing support through a mask interface outside the airway and is used for managing various pediatric disorders that disrupt breathing during sleep (5-7). Home NIV therapy improves the quality of life and well-being of pediatric patients by providing chronic ventilation support, maintaining airway patency, and preventing suboptimal ventilation during sleep (8-10). It also avoids the need for invasive ventilation and reduces respiratory morbidity and mortality (6, 7, 9, 11). Some indications for NIV include chronic respiratory failure, musculoskeletal weakness, obstructive sleep apnea, craniofacial malformations, sleep disorders associated with neurological conditions, and abnormalities in the central respiratory drive (6-8, 10, 12-15).

Providing NIV therapy in the pediatric population requires focused research, as children are unique physiologically (16). For example, pediatric respiratory physiology highly depends on age, weight, and developmental stage, which adds additional challenges related to adherence, including side effects such as nasal symptoms, eye irritation, skin breakdown, mid-face hypoplasia, increased drooling, and gastrointestinal symptoms (6, 11-13, 17). Therefore, healthcare practitioners need an advanced understanding to support and optimize adherence to pediatric NIV therapy, ensuring patients and families receive evidence-informed and personalized care that improves NIV use and overall health for children.

Literature has focused on NIV adherence rates in the pediatric population and described factors associated with poor adherence, including poor mask fit, are partly due to limited options for pediatric interfaces and headgear, as well as the development of short- and long-term complications associated with the therapy (7, 9, 12, 18-20). Complications may be related to inappropriate mask fitting, or the pressure exerted by the interface (6, 17). Factors influencing

adherence to NIV therapy encompass various elements, such as the occurrence of fewer side effects, the type and fitting of the mask, sex, age, maternal education level, developmental delays, baseline apnea-hypopnea index, oxygen saturation level, arousal index level, and sleep maintenance difficulties (6, 18, 19, 22, 41).

Appropriate mask fitting and headgear adaptation are crucial for increasing adherence rates and decreasing the likelihood of complications (9, 11, 22, 23). In 2022, the European Respiratory Society (ERS) published a guideline for NIV initiation in children, emphasizing appropriate mask and headgear fit as vital for optimal initiation and maintenance of NIV (6). Additionally, the 2018 Canadian Pediatric Home Mechanical Ventilation guideline stated that for NIV, interfaces need to be trialed to ensure comfort and effectiveness, which again emphasizes the importance of mask fitting and headgear adaptation in NIV therapy (15). However, to date, original research evaluating strategies to improve mask and headgear fit and, ultimately, improved adherence is limited. This gap identifies the need to research whether interventions to improve mask fit and the timing of these interventions potentially shape outcomes for pediatric patients using NIV therapy.

NIV Initiation and Mask Fitting

Mask selection and well-fitting headgear are essential in the initiation and maintenance of long-term NIV therapy in the pediatric population (11, 17, 25). Five standard mask interface types, including the nasal mask, the oro-nasal mask, the nasal pillow, the total face mask, and the mouthpiece, were available for the pediatric population in the early 2010s (11, 14). Nasal masks are the most commonly used interfaces in the pediatric population (6, 24). Oronasal masks, nasal pillows, and total face masks (only used in intensive care unit settings) may be indicated in cases of poor tolerance or side effects from nasal masks (11, 24). When selecting a mask, it is

important to consider the individual patient's needs and the potential for complications (6, 11, 24). Proper mask fitting is key to minimizing complications and ensuring effective therapy (6, 11, 24).

There have been advancements in pediatric NIV therapy, with case reports of using three-dimensional printing technologies to enhance mask fitting and prevent complications (27-29). However, the current clinical practice remains heavily reliant on the expertise of clinicians in selecting and evaluating optimal masks based on the age of the child and the type of ventilatory support required (8, 9, 11, 42). The limited availability of pediatric masks and headgear during the study period between 2012-2015 made proper mask fitting challenging, especially for children with craniofacial abnormalities and very young infants, leading to increased side effects and reduced adherence (6, 11, 14, 27, 43). Customization becomes imperative to ensure a proper mask fit and mitigate unwanted side effects. Even though there are documented benefits of customization, such as improved adherence to therapy and decreased skin injuries among children (17, 25, 27), there remains a lack of research exploring the customization of headgear and the optimal timing for mask fitting and headgear adaptation. This gap persists despite the recognized impact on adherence rates and the development of complications, as indicated by expert opinion (6).

Adherence to NIV Therapy

NIV compliance reports contain detailed information about the patient’s respiratory cycle and NIV usage (25, 36). The four adherence variables listed in the compliance report include average usage on total days, average usage on days used only, days used, and percent of days used over 4 hours (44). Parameters defining adequate NIV adherence for the pediatric population lack universal validation and are often extrapolated from adult standards (6, 25). In adults,

acceptable overnight NIV adherence typically involves usage for at least four hours a night for a minimum of five nights (70%) per week (8, 14, 25). While this definition is commonly applied in pediatrics, its limitations are acknowledged, particularly in younger children (<14 years) who may have longer sleep durations (32). The primary goal is optimal adherence, so using NIV throughout the entire sleep duration is advocated (6).

Although a standardized definition for NIV adherence rates in the pediatric population is lacking, earlier research emphasizes the predictive value of early adherence patterns for long-term outcomes (7, 18, 19, 37). Consequently, identifying and understanding early adherence patterns can provide valuable insights for healthcare practitioners caring for children requiring long-term NIV.

Complications to NIV Therapy

Improper mask fitting is one of the leading barriers to adherence and a significant cause of complications among infants and children using home NIV therapy (11, 14, 30, 41, 45-47). Interface-related complications are common and can be categorized into several types: mask-related skin issues (e.g., redness to severe skin breakdown and ulcers), short-term pressure-related issues (e.g., bloating and gastric distention), leak-related issues (e.g., eye irritation and mouth leaks), humidification-related issues (e.g., nosebleeds and airway dryness), and long-term complications (e.g., pneumothorax and midface hypoplasia) (11, 12, 14, 16-18, 24, 38, 39).

Many complications, especially mask-related skin issues, are typically related to poor mask fit and are correctable with optimally fitted masks and headgear (11, 14, 16, 25, 38, 39). These potential complications of therapy may be minimized and even eliminated with optimal, customized equipment (11, 14, 16, 25, 38, 39). To our knowledge, no previous research has

captured the impact of mask fitting and headgear customization and when it should be done to minimize complications.

Stollery's Pediatric NIV Program

Pediatric home NIV therapy is a highly specialized field. The Stollery Children's Hospital is a tertiary pediatric medical center with 236 beds located in Alberta, Canada, with a unique, interdisciplinary, and comprehensive pediatric home NIV program (40). The program is led by a nurse practitioner (NP) and joined by a team of pediatric respirologists and respiratory therapists. It is also supported by social workers, occupational therapists (inpatient and outpatient), child life specialists, and orthotists.

A vital aspect of the program is determining the need for individualized mask fitting and headgear adaptation before initiating NIV therapy. The Stollery's mask fit program was established through a recognized need to optimize NIV masks for infants and children when commercial headgear could not support the appropriate use of mask interfaces. Initially, not every patient required mask fitting and headgear adaptation prior to starting NIV therapy at the NIV clinics. However, over the study period, there was a shift towards a more proactive approach of integrating masking fitting and headgear adaptation (if required) before initiating NIV, aiming to enhance the tolerance to and effectiveness of the therapy and reduce adverse effects from available pediatric interfaces from the outset.

Notably, the mask fit and headgear adaptation program at the Stollery is unique and originated from an established need when standard headgear did not fit an infant/child.
As headgear is the key factor in mask stabilization, which adapts to fit a child's unique head shape, it ensures not only comfort but also usability for the child (11). During this process, occupational therapists and occupational therapy assistants thoroughly evaluate the patient's

facial structure and NIV mask fit for a precise fit through adjustment of interface straps, addition of non-skid materials to stabilize the headgear and even utilizing several headgear types to create a new modified headgear that optimizes the mask fit and relieves pressure points (9). Furthermore, custom helmets designed by orthotists are available for highly specialized patients, such as those with plagiocephaly or facial deformities. These helmets accommodate unique cranial shapes and facial traits, enhancing mask fitting and ensuring more effective and comfortable NIV therapy.

This shift towards a proactive mask fitting and headgear adaptation at the Stollery highlights the program's individualized patient care model and commitment to enhancing the NIV adherence rates by optimizing the NIV mask fit. Over time, the team describes that customized headgear optimizes mask fits, which is essential for promoting increased NIV tolerance and use. As a result, the program continues to grow and is utilized today despite the increasing number of pediatric interface options. Currently, no literature on the optimal timing for such interventions is available. The Stollery NIV team theorized that commencing mask fitting early, prior to the initiation of home NIV, could potentially have a positive effect on initial and short-term adherence rates.

Methodology

Study Design

This retrospective case-control study analyzed patient data from the Stollery NIV program between 2012 and 2015. During this period, clinical practice evolved through an identified need to fit infants and children with well-fitting masks. Initially, sleep vendors were responsible for providing the respiratory equipment, and the program would only provide mask fitting and headgear adaptation services after NIV initiation, during the first follow-up clinic visit

within the next three to six months. A more proactive strategy of mask and headgear customization prior to NIV initiation was subsequently established in 2012. This practice change was implemented in the hopes that optimization of mask and headgear fits at an earlier time point would potentially help children initiate and maintain NIV adherence to therapy. To test our hypothesis, NIV adherence data from machine downloads at one, three-, and six months post-NIV initiation were analyzed.

Ethics approval for this project was obtained from the University of Alberta Health Ethics Research Board (Pro00120218).

Population

The study's sample included a pediatric patient cohort that met the following criteria:

1. Age range of 0 to 17 years.
2. Documented visits with the pediatric NIV program at the Stollery Children's Hospital between 2012 and 2015 with the intention of initiating home NIV.
3. Underwent mask and headgear fitting at any point during the study period.

Recruitment

Eligible participants were identified through the program’s patient list, which contains information on patients with documented clinic visits with the pediatric NIV program at the Stollery Children’s Hospital and completed specialized mask fittings between January 1, 2012, and December 31, 2015.

Data Collection

Data for this study were previously collected from the medical charts and NIV machine downloads of patients included in the existing database. Missing data were addressed by cross-referencing original medical charts. Demographics and clinical variables included date of birth,

geographic location of home residence, the primary reason for NIV initiation (conditions that involve upper airway, pulmonary, musculoskeletal, central nervous system, and cardiac systems), location of NIV initiation (inpatient or outpatient), age at NIV initiation, NIV initiation and discontinuation status within six months after NIV recommendation based on machine adherence reports and clinical documentation. Equipment-related information was also extracted, including the type of NIV therapy (CPAP or BIPAP), the type of masks used (nasal or full-face), headgear adaptation status, and numbers of mask fittings within the first six months after NIV recommendation. NIV adherence variables reflected NIV use within 30 days, and data were extracted from the adherence reports at 1, 3, and 6 months from NIV initiation. Appendix C includes all collected variables from research electronic data capture (REDCap), providing a comprehensive overview of the dataset used in this study.

NIV adherence data were obtained directly from local drives of the two most commonly utilized sleep vendors collaborating with the pediatric NIV program and confirmed with the information documented in the medical charts. All of the available machine-generated data were captured during the study period. Prior to the adherence data release, the researchers confirmed appropriate consent forms from patients' guardians.

Outcomes of Interest

Outcomes of interest included:

1. Overall NIV initiation rates within six months from the initial visit with the NIV program as well as at 1, 3 and 6 months.
2. Average nightly hours of NIV use over a 30-day period at 1, 3, and 6 months.
3. Average nightly hours of NIV use on nights used over a 30-day period at 1, 3, and 6 months.
4. Number of nights worn over a 30-day period at 1, 3, and 6 months.

5. Percentage of nights with >4 hours of NIV use over a 30-day period at 1, 3, and 6 months.
6. Discontinuation rate at 6 months from the initial visit with the NIV program.

Database Storage and Access

Study data (including identifying information, clinical information about mask fitting and headgear adaptation, and NIV adherence data) were extracted from previously collected clinical databases, properly stored in a hospital IT drive, and kept confidential. Study data were then uploaded and securely stored in a REDCap database hosted by the Women & Children's Health Research Institute at the University of Alberta (2).

Data Management and Analysis

Once data extraction was complete, data cleansing was performed to ensure accuracy. Data errors were eliminated, and inconsistencies and duplicate entries were removed. All patient data was de-identified prior to analysis. Data analysis was completed using IBM SPSS Statistics version 29 (48).

Participants were first categorized into case and control groups based on the timing of mask fitting in a 2:1 ratio. The **case group** consisted of patients who underwent mask fitting and headgear adaptation (if needed) during the initial visit with the NIV program before starting NIV therapy. The **control group** included patients who had a mask fit and headgear adaptation (if needed) done after NIV initiation. Patients who underwent multiple mask fittings and headgear adaptations throughout the study period to optimize NIV use as they grew and developed were treated as separate entries for adherence analysis as long as they occurred at least three months apart. This approach allowed for the evaluation of the impact of additional fittings on adherence.

Using the Shapiro-Wilk test, a normality analysis was first performed on all collected data. Descriptive statistics were then analyzed to understand the characteristics of the

participants. Given the non-normality indicated by the Shapiro-Wilk test, medians and interquartile ranges (IQR) were calculated for continuous variables and percentages for categorical variables. An alpha level of 0.05 was used for all statistical tests.

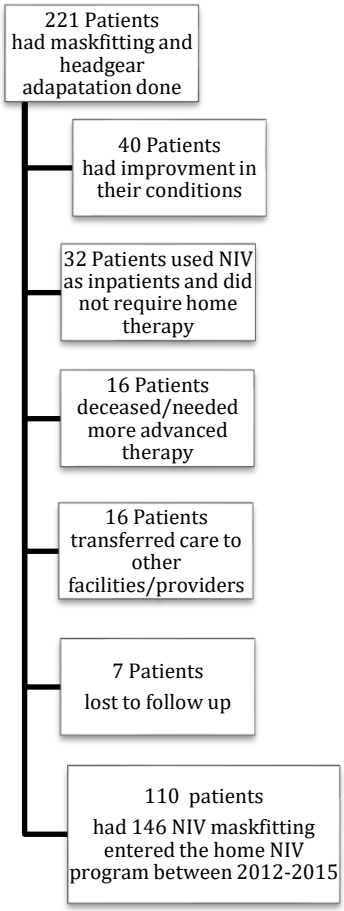
A comparative analysis of demographic and clinical variables between the case and control groups followed the descriptive analysis. The differences between groups were assessed using Pearson's chi-squared test for categorical data and the Mann-Whitney U test for continuous data. Subsequently, Pearson's chi-squared test and odds ratio (OR) with 95% confidence intervals (CI) were used to examine the relationship between early mask fitting and NIV initiation status at 1-, 3-, and 6 months after NIV therapy was recommended.

The median and IQR were utilized as primary statistics for central tendency and variability for comparative analysis of the adherence variables measured at 1-, 3-, and 6-month intervals post-initiation between cases and controls. Due to the non-normality of adherence data, the Mann-Whitney U test was employed to compare the case and control groups. Furthermore, the Kruskal-Wallis H test was conducted separately for the case and control groups to compare adherence rates at 1-, 3-, and 6-month intervals post-initiation, assessing whether there were significant differences in adherence rates within each group over time. Lastly, Pearson's chi-squared test and OR with 95% CI were used to examine the relationship between early mask fitting and NIV discontinuation status at 6 months after NIV therapy was recommended. A comparative analysis was also conducted using Pearson's chi-squared test and Mann-Whitney U tests of demographic and clinical variables between the case and control groups at NIV discontinuation.

Results

During the study period, the Stollery NIV program saw 221 new patients through consultations, either with the intention of starting NIV or in preparation for a titration sleep study where a mask would be needed (Figure 2), accounting for 330 mask-fitting/headgear adaptation occasions. Of these, 111 patients did not meet inclusion criteria: 40 patients did not initiate NIV as their underlying medical condition improved and NIV was no longer required, 32 patients initiated NIV therapy during an acute hospitalization but did not require long-term home NIV therapy upon discharge, 16 patients required a transition to invasive ventilation via tracheostomy or had unfortunately passed away before initiation of home NIV, 16 patients were transferred for care to adult NIV programs or providers in other provinces, and 7 patients were lost to follow-up.

Figure 2. Process of Patients' Exclusion.



Abbreviations: NIV = non-invasive ventilation.

Demographic and Clinical Characteristics of the Study Population

One hundred and ten patients (29% females) met the inclusion criteria of having been mask-fitted through the Stollery pediatric NIV program and provided home equipment to initiate NIV, accounting for 146 patient entries of mask-fitting/headgear adaptation occasions. Close to half of the children reside in the local Edmonton community, the largest city in Northern Alberta, where the Stollery Children's Hospital is located. Around 45% of the children came from suburban, rural, or remote areas. Less than 10% of children came from surrounding provinces served by the Stollery Children's Hospital. Upper airway conditions (64%) emerged as the primary indication for initiating NIV, followed by musculoskeletal conditions (13%), central nervous system disorders (12%), pulmonary conditions (6%), and cardiac conditions (5%). The median age at NIV initiation was 8 years (IQR 1-13), with most patients (72%) starting therapy in outpatient settings, mostly at home. CPAP (63%) was more commonly used, and most patients were fitted with a nasal mask provided by the Stollery NIV clinic (91%). All patients were mask-fitted, either before or after NIV initiation; 27% required headgear adaptations, and 33% required more than one mask fit/headgear adaptation.

Overall, 84 (76%) patients were able to initiate NIV and had clear evidence of NIV use either through adherence download data or clear documentation in the medical chart of NIV being initiated within six months of NIV being recommended, regardless of when they were mask-fitted and whether or not they received a headgear adaptation. When adherence report data was only used to determine NIV initiation, 64 patients were able to initiate, and the NIV initiation rate was 58%. 20 (18%) had no adherence reports available (i.e., SD card malfunction, sleep vendor local drive system upgrades, NIV machines provided by other sleep vendors that

did not provide download reports) but clinical-reported documentation of NIV initiation. These patients were excluded from the comparative analysis since NIV initiation could not be verified with objective NIV usage data. Further details about patient demographics and clinical profiles are depicted in Table 1.

Table 1. Overall Patient Demographics and Clinical Profiles (n = 110).

		N (%)
Sex	Male	78 (71)
	Female	32 (29)
Geographic location	Edmonton (city)	52 (47)
	Within Alberta (rural)	49 (45)
	Out of Alberta	9 (8)
Primary reason for NIV initiation	Upper airway	71 (64)
	Musculoskeletal system	14 (13)
	Central nervous system	13 (12)
	Pulmonary	7 (6)
	Cardiac	5 (5)
Age category at NIV initiation (years old)	< 2	25 (23)
	2-5	17 (15)
	5-13	34 (31)
	>13	34 (31)
Age at NIV initiation	Median (IQR)	8 (1-13)
Initiation location	Outpatient	79 (72)
	Inpatient	31 (28)
NIV therapy type	CPAP	69 (63)
	BIPAP	41 (37)
NIV mask type	Nasal mask	100 (91)
	Full face mask	10 (9)
Headgear adaptation performed	Yes	30 (27)
	No	80 (73)
Need for multiple mask fittings within six months	Yes	36 (33)
	No	74 (67)
NIV initiation status within six months based on clinical documentation	Yes	84 (76)
	No	26 (24)
NIV initiation status within six months based on adherence reports	Yes	64 (58)
	No	46 (42)

Abbreviations: NIV = non-invasive ventilation, IQR = interquartile range, CPAP = continuous positive airway pressure, BIPAP = bilevel positive airway pressure.

Comparison of Demographic and Clinical Characteristics of Cases and Controls

76 patients (69%) had mask fit/headgear adaptation services prior to initiating NIV (cases), while 34 patients (31%) started NIV with the standard equipment provided by the sleep vendors and were mask fit/headgear adapted during one of the follow-up visits with the NIV program (controls) within the first six months. Overall, the case and control groups were homogeneous in terms of their demographic and clinical characteristics as well as equipment-related characteristics, with no significant differences noticed between groups. Of note, the variable "age at initiation" was also tested. As indicated, the median age at initiation for the case group was 9 years old (IQR 3-13), and for the control group, it was 6 years old (IQR -12), with no overall significant differences (p=0.256). There were also no significant differences between the groups when comparing mask fit and headgear adaptation. Specifically, 17 patients (22%) in the case group and 13 patients (38%) in the control group required headgear adaptation, with no statistically significant difference (p=0.084). Additionally, 25 patients (33%) in the case group and 11 patients (32%) in the control group required multiple mask fittings within the first six months, also showing no significant difference (p=0.955). Table 2 depicts further details about the comparative analysis of clinical and demographic variables at baseline.

Table 2. Comparative Analysis of Clinical and Demographic Variables at Baseline (n =110).

Demographic and clinical variables		Cases N (%)	Controls N (%)	Chi-square	df	P
Total number of participants		76 (100)	34 (100)			
Geographic location	Edmonton (city)	36 (47)	16 (47)	0.027	2	0.986
	Within Alberta (rural)	34 (45)	15 (46)			
	Other provinces	6 (8)	3 (7)			
Sex	Female	20 (26)	12 (35)	.918	1	0.338

Primary reason for NIV initiation	Upper airway	48 (63)	23 (67)	1.903	4	0.754
	Musculoskeletal system	9 (12)	5 (15)			
	Central nervous system	11 (14)	2 (6)			
	Pulmonary	5 (7)	2 (6)			
	Cardiac	3 (4)	2 (6)			
Age category at NIV initiation (years old)	< 2	13 (17)	12 (35)	5.720	3	0.126
	2-5	14 (19)	3 (9)			
	5-13	23 (30)	11 (32)			
	>13	26 (34)	8 (24)			
Age at NIV initiation	Median (IQR)	9 (3-13)	6 (1-12)			0.256
Initiation location	Outpatient	57 (75)	22 (65)	1.230	1	0.267
	Inpatient	19 (25)	12 (35)			
NIV therapy type	CPAP	48 (63)	21 (60)	0.020	1	0.889
	BIPAP	28 (37)	13 (40)			
NIV mask type	Nasal mask	69 (91)	31 (91)	0.04	1	0.948
	Full face mask	7 (9)	3 (9)			
Need for headgear adaptation	Yes	17 (22)	13 (38)	7.194	1	0.084
Need for multiple mask fitting within six months	Yes	25 (33)	11 (32)	0.003	1	0.955

Abbreviations: df = degree of freedom, NIV = non-invasive ventilation, IQR = interquartile range, CPAP = continuous positive airway pressure, BIPAP = bilevel positive airway pressure.

Comparison of NIV Initiation Rates between Cases and Controls

Overall, 46 children (61%) in the case group had evidence of NIV use, compared to 18 children (53%) in the control group (p=0.456) with an OR of 1.36 (95% CI: 0.60 to 3.08). There were no significant differences in initiation rates within the first 6 months since the initial visit with the NIV program between patients who underwent mask fitting and headgear adaptation prior to initiating NIV (cases) and those who received these interventions following NIV initiation (controls) based on adherence download data.

At one month following the initial visit with the NIV program, 41 children (54%) in the case group had evidence of NIV use in available downloads, and 15 children (44%) in the

control group. This difference was not statistically significant (p = 0.341), with an OR of 1.48 (95% CI: 0.66 to 3.35). By three months, 43 children (57%) in the case group had evidence of NIV use, compared to 17 (50%) in the control group, again showing no significant differences (p=0.522) with an OR of 1.30 (95% CI: 0.58 to 2.93). By six months, 46 children (61%) in the case group had evidence of NIV use, compared to 18 children (53%) in the control group. This difference remained non-significant (p = 0.456), with an OR of 1.36 (95% CI: 0.60 to 3.08). Further details are provided in Table 3.

Table 3. Documented NIV Initiation at 1-, 3-, and 6-months after NIV Recommendation (n=110).

NIV Initiation rate	Cases N (%)	Controls N (%)	Chi-square	df	P	OR (OR 95% CI)
Total number of participants	76 (100)	34 (100)				
NIV initiation status within one month based on adherence report	41 (54)	15 (44)	0.908	1	0.341	1.48 (0.66 to 3.35)
NIV initiation status within three months based on adherence report	43 (57)	17 (50)	0.410	1	0.522	1.30 (0.58 to 2.93)
NIV initiation status within six months based on adherence report	46 (61)	18 (53)	0.555	1	0.456	1.36 (0.60 to 3.08)

Abbreviations: NIV = non-invasive ventilation, df = degree of freedom, OR = odds ratio, CI = confidence intervals.

Comparative Analysis of NIV Adherence at 1-, 3-, and 6-Months post NIV Initiation

All of the 73 available adherence reports were obtained from the sleep vendors, providing a comprehensive dataset for analyzing the adherence patterns over time between the cases and controls. There were 43 adherence reports in the case group and 20 adherence reports available in the control group 1 month after the initial NIV program visit. At 3 months, 36 adherence reports were available for the cases and 18 for the controls. By 6 months, 32 adherence reports

were available for the case group, and 14 were available for the control group. Overall, no significant differences in NIV adherence parameters were observed between patients who were mask-fitted before NIV initiation (case group) and those who have fitted afterward (control group) at 1-, 3-, and 6 months after NIV initiation.

At 1 month, the median nightly NIV usage over a 30-day period was 273 minutes (IQR 32-486) in the case group compared to 243 minutes (IQR 19-333) in the control group, with no significant difference (p=0.4). At 3 months, the cases' median nightly usage was 344 minutes (IQR 88-565) compared to 257 minutes (IQR 27-419) in the controls (p=0.13). By 6 months, the median nightly NIV usage was 377 minutes (IQR 34-561) in the case group versus 308 minutes (IQR 52-423) in the control group (p=0.51).

When examining NIV usage on only the days it was used, at 1 month, the median usage in the case group was 347 minutes (IQR 152-513) compared to 313 minutes (IQR 75-392) in the control group (p=0.55). At 3 months, the case group had a median usage of 391 minutes (IQR 147-578) compared to 312 minutes (IQR 87-467) in the control group (p=0.37). By 6 months, the case group's median usage on days used increased to 476 minutes (IQR 109-567) compared to 331 minutes (IQR 105-528) in the control group (p=0.29).

For the number of days with NIV use, at 1 month, the case group had a median of 18 days (IQR 7-29) of use compared to 19 days (IQR 6-30) in the control group (p=0.67). At 3 months, the case group had a median of 22 days (IQR 13-30) of NIV use compared to 17 days (IQR 2-28) in the control group (p=0.29). By 6 months, the case group had a median of 27 days (IQR 14-30) of NIV use compared to 24 days (IQR 7-30) in the control group (p=0.43).

Lastly, when examining the percentage of days with more than 4 hours of NIV use, at 1 month, 56% (IQR 7-96) of days in the case group had more than 4 hours of use, compared to

45% (IQR 2-68) in the control group (p=0.21). At 3 months, the case group had 60% (IQR 5-100) of days with more than 4 hours of use, compared to 46% (IQR 2-89) in the control group (p=0.34). By 6 months, 88% (IQR 1-100) of days in the case group had more than 4 hours of NIV use, compared to 61% (IQR 4-84) in the control group (p=0.18). Tables 4, 5, and 6 depict further details regarding comparative analysis.

Table 4. Adherence Variables Comparison of Case/control Groups at 1-month (n=63).

NIV adherence at 1 month (30-day period)	Groups	N	Median	IQR	Mean rank	Sum of ranks	U	P
Average NIV usage on total days (in mins)	Case	43	273	32-486	29.18	1432.5	373.5	0.4
	Control	20	243	19-333	33.31	583.5		
Average usage on days used only (in mins)	Case	43	347	152-513	29.1	1434	372	0.39
	Control	20	313	75-392	33.35	582		
Days used	Case	43	18	7-29	31.58	1358	412	0.79
	Control	20	19	6-30	32.9	658		
Percent of days use over 4 hours (%)	Case	43	56	7-96	33.66	1447.5	358.5	0.29
	Control	20	45	2-68	28.43	568.5		

Abbreviations: NIV = non-invasive ventilation, IQR = interquartile range.

Table 5. Adherence Variables Comparison of Case/control Groups at 3-month (n=54).

NIV adherence at 3 months (30-day period)	Groups	N	Median	IQR	Mean rank	Sum of ranks	U	P
Average usage on total days (in mins)	Case	36	344	88-565	29.79	1072.5	241.5	0.13
	Control	18	257	27-419	22.92	412.5		
Average usage on days used only (in mins)	Case	36	391	147-578	29.79	1072.5	241.5	0.13
	Control	18	312	87-467	22.92	412.5		
Days used	Case	36	22	13-30	30.33	1092	222	0.06
	Control	18	17	2-28	21.83	393		
Percent of days use over 4 hours (%)	Case	36	60	5-100	29.35	1056.5	257.5	0.22
	Control	18	46	2-89	23.81	428.5		

Abbreviations: NIV = non-invasive ventilation, IQR = interquartile range.

Table 6. Adherence Variables Comparison of Case/control Groups at 6-month (n =46).

NIV adherence at 6 months (30-day period)	Groups	N	Median	IQR	Mean rank	Sum of ranks	U	P
Average usage on total days (in mins)	Case	32	377	34-561	24.36	779.5	196.5	0.51
	Control	14	308	52-423	21.54	301.5		
Average usage on days used only (in mins)	Case	32	476	109-567	24.36	779.5	196.5	0.51
	Control	14	331	105-528	21.54	301.5		
Days used	Case	32	27	14-30	24.08	770.5	205.5	0.65
	Control	14	24	7-30	22.18	310.5		
Percent of days use over 4 hours (%)	Case	32	88	1-100	25.22	807	169	0.18
	Control	14	61	4-84	19.57	274		

Abbreviations: NIV = non-invasive ventilation, IQR = interquartile range.

Changes in Adherence Rates Over Time within the Case and Control Groups

Available adherence data at 1, 3, and 6 months were compared within both the case and control groups to determine changes over time. Overall, NIV usage remained consistent over time during the first six months after NIV initiation in both case and control groups. Tables 7 and 8 provide further details.

Table 7. Case group's Adherence Rates at 1-, 3-, and 6-month Intervals Post-initiation.

NIV adherence variables	Time point	Cases (N)	Mean rank	Kruskal-Wallis H	df	P
Average usage on total days	1-month	43	50.58	2.001	2	0.368
	3-month	36	59.82			
	6-month	32	58.98			
Average usage on days used only	1-month	43	51.55	1.367	2	0.505
	3-month	36	59.38			
	6-month	32	58.19			
Days used	1-month	43	49.42	3.047	2	0.218
	3-month	36	60.85			
	6-month	32	59.39			
Percent of days use over 4 hours	1-month	43	52.88	0.726	2	0.695
	3-month	36	57.18			
	6-month	32	58.86			

Abbreviations: NIV = non-invasive ventilation, df = degree of freedom.

Table 8. Control Group's Adherence Rates at 1-, 3-, and 6-month Intervals Post-initiation.

NIV adherence variables	Time point	Controls (N)	Mean rank	Kruskal-Wallis H	df	P
Average usage on total days	1-month	20	25.05	0.493	2	0.781
	3-month	18	26.36			
	6-month	14	28.71			
Average usage on days used only	1-month	20	26.25	0.086	2	0.958
	3-month	18	26.00			
	6-month	14	27.50			
Days used	1-month	20	27.73	0.964	2	0.618
	3-month	18	23.69			
	6-month	14	28.36			
Percent of days use over 4 hours	1-month	20	25.73	0.090	2	0.956
	3-month	18	26.83			
	6-month	14	27.18			

Abbreviations: NIV = non-invasive ventilation, df = degree of freedom.

Comparison of Discontinuation Rates between Cases and Controls at 6 Months

Out of the 64 patients who had adherence reports during the study period, 11 (17%) had a documented discontinuation of NIV within six months after initiation. When examining the dropout rates between case and control groups, 7 patients (15%) in the case group and 4 patients (22%) in the control group stopped using NIV therapy within the first 6 months after initiation (p=0.504) with an OR of 0.63 (95% CI: 0.16 to 2.48). The demographic and clinical characteristics of patients who dropped between cases and controls were similar (Table 9).

Notably, the median age at initiation for the case group was 5 years (IQR 3-15), and for the control group, it was 2 years (IQR 0-12). However, the two groups had similar dispersion measures and no overall significant differences (p=0.257). There were also no significant differences between the groups when comparing mask fit and headgear adaptation. Specifically, 1 patient (14%) in the case group and 1 patient (25%) in the control group required headgear adaptation, with no statistically significant difference (p=0.658). Additionally, 4 patients (57%)

in the case group and 3 patients (75%) in the control group required multiple mask fittings, also showing no significant difference (p=0.554).

Table 9. Comparative Analysis of Clinical and Demographic Variables who Discontinued at 6 Months (n = 11).

Demographic and clinical variables		Cases N (%)	Controls N (%)	Chi-Square	df	P
Patients who dropped out	OR (95% CI) 0.63 (0.16 to 2.48)	7 (100)	4 (100)	0.446	1	0.504
Geographic location	Edmonton (city)	2 (29)	1 (25)	0.016	1	0.898
	Within Alberta (rural)	5 (71)	3 (75)			
	Out of province	0 (0)	0 (0)			
Sex	Female	2 (28)	1 (25)	0.016	1	0.898
Primary reason for NIV initiation	Upper airway	3 (43)	2 (50)	5.814	4	0.213
	Musculoskeletal system	0 (0)	2 (50)			
	Central nervous system	1 (14)	0 (0)			
	Pulmonary	1 (14)	0 (0)			
	Cardiac	2 (29)	0 (0)			
Age category at NIV initiation (years old)	< 2	1 (14)	2 (50)	1.997	3	0.573
	2-5	2 (29)	1 (25)			
	5-13	1 (14)	0 (0)			
	>13	3 (43)	1 (25)			
Age at NIV initiation	Median (IQR)	5 (3-15)	2 (0-12)			0.257
Initiation location	Outpatient	5 (71)	2 (50)	0.505	1	0.477
	Inpatient	2 (29)	2 (50)			
NIV therapy type	CPAP	4 (57)	2 (50)	0.052	1	0.819
	BIPAP	3 (43)	2 (50)			
NIV mask type	Nasal mask	7 (100)	4 (100)			
	Full face mask	0 (0)	0 (0)			
Need for headgear adaptation	Yes	1 (14)	1 (25)	0.196	1	0.658
Need for multiple mask fitting within six months	Yes	4 (57)	3 (75)	0.351	1	0.554

Abbreviations: df = degree of freedom, NIV = non-invasive ventilation, IQR = interquartile range, CPAP = continuous positive airway pressure, BIPAP = bilevel positive airway pressure.

Discussion

While the current understanding of pediatric home NIV therapy highlights factors such as patient comfort, medical condition severity, and family involvement as some of the key influencers of adherence, our study found that the timing of mask fitting did not significantly impact NIV initiation, adherence, or discontinuation (6, 11, 16, 18, 46, 49-51). This suggests that adherence may be influenced by a more complex interplay of factors, with elements beyond the timing of mask fitting potentially playing a more crucial role in successful NIV therapy. Notably, adherence rates in both groups in our study were higher than previously reported in the literature, suggesting that mask fitting and headgear adaptation—whether performed prior to or shortly after NIV initiation—are key interventions for successful therapy, which are consistent with the current guideline recommendations (6, 15). Although the timing of mask fitting in this study did not influence NIV initiation, adherence or discontinuation, our findings offer valuable insights into pediatric NIV therapy adherence from a different perspective.

Role of Mask and Headgear Fitting in NIV Initiation and Adherence

Choosing an adequate, properly fitting interface is believed to be imperative to successfully support infants and children requiring NIV (6, 11, 15, 17, 24, 25, 47, 52). Although this study could not determine whether the timing of mask and headgear fitting prior to NIV initiation could facilitate NIV initiation and adherence, in our clinical practice, our experience is that it is an essential part of optimizing NIV use and care of these patients. Further, we observed overall higher adherence rates within the first six months post-NIV initiation in our cohort in comparison to current literature (14, 21, 45, 46, 53-56). A meta-analysis by Sawunyavisuth et al. (2022) reported an adherence rate of 46.53% (9,883/21,240) across 13 studies based on the percentage of days used for ≥ 4 hours (21). Pascoe et al. (2019) found that 42 youths with

Duchenne muscular dystrophy had adherence rates of 56.1% ± 38.7% for days used and 46.2% ± 40.6% for days used ≥ 4 hours, with an average usage of 5.61 ± 4.23 hours on days worn (46). Ennis et al. (2015) reported a mean BiPAP usage of 6.04 ± 3.47 hours per night, while Marcus et al. (2012) noted a mean NIV usage of 170 ± 145 minutes per night (45, 56). Simon et al. (2012) observed an average NIV usage of 3.35 ± 2.79 hours per night and 5.01 ± 2.51 hours per night on days used, with 41% ± 35% of days used for ≥ 4 hours (55).

Prior to or within the first six months after NIV initiation, Stollery's standard of care is the provision of a mask fitting/customization of headgear by a highly specialized multidisciplinary team, which might explain the overall higher initiation rates and adherence. It is reasonable to consider that providing these services before NIV initiation might prevent negative early experiences with inadequate fit, thereby reducing resistance from patients and families using NIV.

Other factors beyond the timing of mask fitting, however, may have also played critical roles in impacting the ability of a child and family to initiate and adhere to NIV, limiting our ability to demonstrate significant differences in adherence rates based on the timing for which mask fit/headgear adaptation services were offered. Previously reported factors included patient comfort, the severity of medical conditions, and the level of family involvement (6, 11, 16, 18, 46, 49-51). Using a patient-centered care model, as an experienced team, the NIV program at the Stollery Children's Hospital actively identifies and addresses these modifiable factors, supports families who have already shown interest in initiating NIV, and acts on modifiable factors (57). This practice affects both case and control groups, minimizing the chances of detecting significant differences.

Maintained NIV Adherence within 6 Months after NIV Initiation

As revealed by the Kruskal-Wallis H test, the maintained adherence rates over time for both the case and control groups underscore several critical factors that might have contributed to this outcome. We hypothesized that early mask fit/headgear prior to NIV initiation might result in earlier initiation and higher NIV usage, which this study could not demonstrate. This was not entirely surprising since our team provides early follow-up during the first one to six months of NIV initiation and remains available for families to call with concerns about their children's equipment. In fact, all children in the control group had an assessment of their mask fit within the first six months, and, in a high proportion of them, their headgear was adapted, correcting potential improper fits from standard equipment. This standard approach, consistent with recommendations from the literature, might have played a pivotal role in sustaining adherence in both cases and control groups and, therefore, limiting our capacity to find differences between groups (6, 11, 14, 17, 24, 25, 30, 41, 45-47). Consistent and proper fitting of the mask and headgear ensures comfort and the effective delivery of NIV therapy, which is essential for promoting continued use among pediatric patients (6, 11, 17, 24, 25). Properly fitted masks and headgear help minimize side effects such as skin irritation, pressure sores, and discomfort, all of which can significantly impact adherence (11, 14, 30, 41, 45-47). The significance of maintaining a proper fit throughout therapy duration cannot be overstated, as discomfort or poor fit can lead to discontinuation or improper use of the therapy, ultimately affecting its efficacy (6, 11, 17, 24, 25).

Secondly, incorporating an interdisciplinary healthcare team approach is crucial in maintaining high adherence rates (6, 15, 54). As the lead of the NIV program, the NP coordinates the overall care plan, ensures that each patient's treatment is tailored to their specific needs, and

monitors their progress throughout the therapy. The respiratory therapists are responsible for participation in the mask fitting, educating patients and families on NIV therapy, and making ongoing adjustments to optimize ventilation support. The social workers offer financial resources to families so patients can receive home NIV therapy without adding extra financial burdens. Occupational therapists and occupational therapy assistants thoroughly evaluate the patient's facial structure and provide headgear adaptation to optimize the mask fit, comfort, and adherence. Child life specialists help children cope with their NIV by using therapeutic play and providing emotional support to reduce anxiety and improve children's ability to adjust to starting NIV therapy and their experiences with NIV therapy over time. Orthotists design and customize helmets for highly specialized patients, such as those with plagiocephaly or scaphocephaly, ensuring their unique cranial shapes are accommodated for effective NIV therapy.

The multidisciplinary approach at Stollery's NIV program provides holistic support to families and may have contributed to the high adherence rates observed in both groups in this study. Since other known factors—such as patient and family education about NIV, social support, familiarity with NIV, and continuous support from the healthcare team—can significantly impact adherence, the comprehensive care offered by the entire team may have enhanced adherence beyond the effects of mask fitting and headgear adaptation alone (6, 11, 16, 18, 21, 46, 49-51). This could explain why no significant differences in adherence were seen between the groups, as both benefited from the robust support provided by the interdisciplinary team. Therefore, while mask fitting and headgear adaptation are important, the broader, patient-centered care approach may also play a significant role in sustaining high adherence rates across all patients in the NIV program.

NIV Discontinuation within 6 Months after NIV Initiation

In this study, there was no statistical significance between the discontinuation rates in the case group and the control group. Overall, the discontinuation rate in the study cohort remained relatively low compared to the literature observed. For example, Guilleminault et al. (1995) reported a 39% discontinuation and a 10% loss to follow-up rate in 72 infants successfully treated at home with CPAP, mostly for reasons including improvement after upper airway surgeries and improvement of polygraphic monitoring results with age (58). Markström et al. (2008) noted a 33% discontinuation rate in 18 infants treated with NIV for reasons including death, improvement in condition, and requiring invasive ventilation (59). McNamara et al. (1999) reported 6 (25%) patients discontinued NIV out of a group of 24 infants; 3 (13%) patients did not use NIV due to family refusal (60). The other families of 3 (13%) infants were not able to manage the NIV at home (60). These differences may suggest that interventions such as ensuring proper mask fitting within the first six months of NIV initiation, implemented in our program, may be effective in promoting and maintaining adherence to NIV therapy. Ensuring a properly fitted mask and headgear throughout therapy, along with frequent follow-up, can help in the early detection and resolution of discomfort or complications that might lead to discontinuation (6, 11, 17, 24, 25). As this is a secondary data analysis focused on the timing of mask fitting's effect on NIV adherence metrics, it is important to note that our study did not capture the depth of detail to analyze the reasons for discontinuation (e.g., mortality, patient/family decisions to stop, improvement in clinical condition, or being lost to follow-up) between the cases and controls. These reasons for discontinuation could have provided deeper insights into the factors influencing adherence and dropout but would fall outside the scope of this research, which is

centered on adherence behaviors rather than the clinical or personal factors driving discontinuation.

Study Limitations

As this was a single-centered and retrospective study based on previously collected data, there are some limitations, and generalizations must be made with caution. Although we had access to all objective adherence data directly available through machine downloads from the datasets of sleep companies, cloud-based systems were not available during the study period. Adherence data was only uploaded to the database if patients and families brought secure digital (SD) memory cards to the sleep companies for uploads, which our NIV program requested for all our families. Unforeseeable reasons, such as company system upgrades or defective SD cards, might have also resulted in some missing data, as highlighted in the results. However, potential missing data should have affected cases and controls equally. This is likely the case since the comparison analysis, including children with documentation of NIV initiation in the medical chart and no adherence reports available, provided similar results. Additionally, a minority of patients were served by sleep companies with no local datasets available; thus, their adherence data was only available if documented in the medical chart. Similarly, this missing data would have likely affected cases and controls in the same way.

Conclusion

The NP-led NIV program at Stollery Children's Hospital has a multidisciplinary team that collaborates with patients and families with the goal in mind: every infant or child enrolled in the NIV program is ensured a designated mask and headgear fitting at any given time while on NIV therapy. From a clinical perspective, early individualized mask fitting with headgear adaptation (if required) has resulted in more effective use and optimization of adherence to

therapy in our Stollery program. This thesis investigated whether the timing of these mask-fitting services prior to NIV initiation affected NIV initiation and adherence rates in the pediatric population.

While this study did not demonstrate a statistically significant impact regarding the timing of mask fitting on NIV initiation and therapy adherence at six months; this study revealed that adherence rates in both the case and control groups were higher than those reported in the literature. This finding suggests that mask fitting and headgear adaptation are essential interventions for successful NIV initiation, whether conducted before or shortly after therapy begins. The comprehensive service offered by specialized pediatric NIV programs is vital in maintaining high adherence rates, ultimately leading to improved health outcomes and well-being for pediatric patients.

Chapter 3: General Discussion and Conclusions

Implications for Future Study

The NIV adherence rates observed in this study aligned with or exceeded those reported in the current literature. Although our results could not demonstrate that a proactive approach to mask fitting and headgear adaptation by a specialized team prior to NIV initiation resulted in a higher likelihood of initiating and adhering to NIV, the overall high adherence rates in both groups potentially reflect the known benefits of a comprehensive multidisciplinary pediatric NIV program. Since mask fitting and headgear adaptation have become the standard of care at the Stollery Children's Hospital, we were not able to collect more data from control groups where patients did not have access to these interventions for additional research.

Stollery's specialized NIV program aims to target previously identified barriers to NIV initiation and adherence. For future studies, engaging pediatric patients using home NIV and their families in the co-design of specific interventions to address identified barriers with NIV use would be important. Through a qualitative participatory approach that identifies barriers to and potential solutions with respect to initiating NIV and using it over time would be beneficial as these children and their families are the ones with the lived experiences with respect to the challenges of initiating and maintaining NIV use. Subsequent research could then implement the interventions recommended and measure their effectiveness, which could serve as a foundation for developing targeted strategies to enhance the efficacy of home NIV therapy and support a new establishment of standardized pediatric home NIV care tailored to Stollery patients. Ultimately, this could improve the overall health outcomes for pediatric patients using home NIV at the Stollery.

Implications for Nursing

The findings from our study suggest that though mask fitting plays a vital role in pediatric NIV therapy, the role of *timing* of mask fitting remains inconclusive. A multidisciplinary and comprehensive approach to NIV therapy was found to be beneficial to achieving an overall higher adherence rate than has been previously shown in the literature. One of the most important strategies that has been found to be a key element in facilitating NIV use in infants and children is the provision of specialized mask fitting and headgear adaptation either before or shortly after NIV initiation, as is the current practice by a specialized team for all infants and children requiring home NIV therapy through the Stollery NIV program. Other individualized strategies may include patient and family education, continuous support, regular follow-up, and ongoing reassessment of equipment and mask fit (6, 50). Addressing barriers to adherence, such as short- and long-term side effects, discomfort, and lack of access to healthcare, is also necessary for long-term NIV adherence (6, 45, 46). These findings are important for healthcare professionals to consider when caring for pediatric patients on home NIV therapy. Decision-makers, such as patient care managers who oversee the allocation of resources, should be informed on the positive effects of these strategies so they can prioritize appropriate resource allocation that can continue to support the ongoing requirement for an NIV mask fitting and headgear adaptation program for Stollery patients requiring NIV therapy. Nurses and nurse practitioners could advocate for pediatric patients to receive specialized NIV programs and care to ensure optimal outcomes. Nursing staff and other frontline clinicians should also stay informed with respect to NIV advanced technologies that could be used for pediatric patients requiring NIV therapy. These could significantly shape future clinical practices and aid in establishing improved long-term therapy routines for this vulnerable pediatric population.

Conclusion

As one of the first studies to explore the impact of the timing of mask fitting on NIV therapy adherence in a pediatric population, our study suggests that early mask fitting/headgear adaptation potentially contributed to overall high adherence rates in our study cohort. However, we could not demonstrate whether offering this specialized care prior to NIV initiation could have resulted in an even larger effect on NIV initiation, maintenance of adherence at 1,3, and 6 months or discontinuation at 6 months after NIV initiation.

Our findings emphasize the importance of an interdisciplinary team and a patient-centered approach in optimizing the initiation and ongoing management of pediatric NIV therapy. Care teams that facilitate NIV initiation, optimize adherence, and ensure positive therapeutic outcomes are essential. Consideration of factors such as patient comfort, caregiver support, and individualized strategies require study. The comprehensive care provided by specialized pediatric NIV programs is vital in addressing the challenges to NIV adherence in infants and children and in maintaining high adherence rates, that will ultimately improve the health outcomes and overall well-being of infants and children requiring home NIV therapy.

References

1. Harris PA, Taylor R, Minor BL, Elliott V, Fernandez M, O'Neal L, et al. The REDCap consortium: Building an international community of software platform partners. J Biomed Inform. 2019;95:103208.
2. Harris PA, Taylor R, Thielke R, Payne J, Gonzalez N, Conde JG. Research electronic data capture (REDCap)--a metadata-driven methodology and workflow process for providing translational research informatics support. J Biomed Inform. 2009;42(2):377-81.
3. Sleep Medix. Sleep Medix, Sleep Apnea Solution 2024 [Available from: https://sleepmedix.com/.
4. Medigas. Medigas, A Lined Company 2024 [Available from: https://www.medigas.com/en.
5. Fauroux B, Khirani S, Griffon L, Teng T, Lanzeray A, Amaddeo A. Non-invasive Ventilation in Children With Neuromuscular Disease. Frontiers in Pediatrics. 2020;8.
6. Fauroux B, Abel F, Amaddeo A, Bignamini E, Chan E, Corel L, et al. ERS statement on paediatric long-term noninvasive respiratory support. European Respiratory Journal. 2022;59(6):2101404.
7. Castro-Codesal ML, Dehaan K, Featherstone R, Bedi PK, Martinez Carrasco C, Katz SL, et al. Long-term non-invasive ventilation therapies in children: A scoping review. Sleep medicine reviews. 2018;37:148-58.
8. Chawla J, Edwards EA, Griffiths AL, Nixon GM, Suresh S, Twiss J, et al. Ventilatory support at home for children: A joint position paper from the Thoracic Society of Australia and New Zealand/Australasian Sleep Association. Respirology. 2021;26(10):920-37.
9. Djkowich M, Olmstead D, Castro-Codesal ML, Scott S. Who is using noninvasive ventilation? A descriptive study examining the population enrolled in a pediatric noninvasive ventilation program. Journal for Specialists in Pediatric Nursing. 2021;26(3).
10. Praud J-P. Long-Term Non-invasive Ventilation in Children: Current Use, Indications, and Contraindications. Frontiers in Pediatrics. 2020;8.
11. Castro-Codesal ML, Olmstead DL, MacLean JE. Mask interfaces for home non-invasive ventilation in infants and children. Paediatric respiratory reviews. 2019;32:66-72.
12. Bedi PK, Castro-Codesal ML, Featherstone R, AlBalawi MM, Alkhaledi B, Kozyrskyj AL, et al. Long-term non-invasive ventilation in infants: A systematic review and meta-analysis. Frontiers in Pediatrics. 2018;6.
13. Castro-Codesal ML, Dehaan K, Bedi PK, Bendiak GN, Schmalz L, Rosychuk RJ, et al. Long-term benefits in sleep, breathing and growth and changes in adherence and complications in children using noninvasive ventilation. Canadian Journal of Respiratory, Critical Care, and Sleep Medicine. 2020;4(2):115-23.
14. Fedor KL. Noninvasive Respiratory Support in Infants and Children. Respir Care. 2017;62(6):699-717.
15. Amin R, MacLusky I, Zielinski D, Adderley R, Carnevale F, Chiang J, et al. Pediatric home mechanical ventilation: A Canadian Thoracic Society clinical practice guideline executive summary. Canadian Journal of Respiratory, Critical Care, and Sleep Medicine. 2017;1(1):7-36.
16. Haut C. Pediatric Noninvasive Ventilation. Journal of pediatric intensive care. 2015;04(02):121-7.

17. Bedi PK, DeHaan K, Ofosu D, Olmstead D, MacLean JE, Castro-Codesal M. Predictors of NIV-related adverse events in children using long-term noninvasive ventilation. Pediatric pulmonology. 2023;58(12):3549-59.
18. Blinder H, Momoli F, Bokhaut J, Bacal V, Goldberg R, Radhakrishnan D, et al. Predictors of adherence to positive airway pressure therapy in children: a systematic review and meta-analysis. Sleep medicine. 2020;69:19-33.
19. Blinder H, Momoli F, Holland SH, Blinder A, Radhakrishnan D, Katz SL. Clinical predictors of nonadherence to positive airway pressure therapy in children: a retrospective cohort study. Journal of clinical sleep medicine : JCSM : official publication of the American Academy of Sleep Medicine. 2021;17(6):1183-92.
20. Praud JP. Long-Term Non-invasive Ventilation in Children: Current Use, Indications, and Contraindications. Front Pediatr. 2020;8:584334.
21. Sawunyavisuth B, Ngamjarus C, Sawanyawisuth K. Adherence to Continuous Positive Airway Pressure Therapy in Pediatric Patients with Obstructive Sleep Apnea: A Meta-Analysis. Ther Clin Risk Manag. 2023;19:143-62.
22. Amaddeo A, Frapin A, Fauroux B. Long-term non-invasive ventilation in children. The Lancet Respiratory Medicine. 2016;4(12):999-1008.
23. Tran T, Nonoyama M, Cithiravel N, Syed F, Janevski J, Chiang J, et al. Virtual mask fitting in pediatric patients during COVID-19: A case series. Canadian journal of respiratory therapy : CJRT = Revue canadienne de la therapie respiratoire : RCTR. 2021;57:93-8.
24. Khirani S, Ducrot V. Mask interfaces and devices for home noninvasive ventilation in children. Pediatric pulmonology. 2024;59(6):1528-40.
25. Choi P, Adam V, Zielinski D. Noninvasive Ventilation Downloads and Monitoring. Sleep medicine clinics. 2020;15(4):569-79.
26. Pigmans RRWP, Klein-Blommert R, van Gestel MC, Markhorst DG, Hammond P, Boomsma P, et al. Development of personalized non-invasive ventilation masks for critically ill children: a bench study. Intensive Care Medicine Experimental. 2024;12(1):21.
27. Borràs-Novell C, Causapié MG, Murcia M, Djian D, García-Algar Ó. Development of a 3D Individualized Mask for Neonatal Non-Invasive Ventilation. Int J Bioprint. 2022;8(2):516.
28. Duong K, Glover J, Perry AC, Olmstead D, Ungrin M, Colarusso P, et al. Feasibility of three-dimensional facial imaging and printing for producing customised nasal masks for continuous positive airway pressure. ERJ Open Research. 2021;7(1):00632-2020.
29. Willox M, Metherall P, Jeays-Ward K, McCarthy AD, Barker N, Reed H, et al. Custom-made 3D printed masks for children using non-invasive ventilation: a feasibility study of production method and testing of outcomes in adult volunteers. J Med Eng Technol. 2020;44(5):213-23.
30. Bockstedte M, Xepapadeas AB, Spintzyk S, Poets CF, Koos B, Aretxabaleta M. Development of Personalized Non-Invasive Ventilation Interfaces for Neonatal and Pediatric Application Using Additive Manufacturing. J Pers Med. 2022;12(4).
31. Brill A-K. How to avoid interface problems in acute noninvasive ventilation. Breathe. 2014;10(3):230-42.
32. Paruthi S, Brooks LJ, D'Ambrosio C, Hall WA, Kotagal S, Lloyd RM, et al. Recommended Amount of Sleep for Pediatric Populations: A Consensus Statement of the American Academy of Sleep Medicine. Journal of clinical sleep medicine : JCSM : official publication of the American Academy of Sleep Medicine. 2016;12(6):785-6.

33. ResMed. myAir app 2024 [Available from: https://support.resmed.com/en-us/digital-health-solutions/myair-app/.
34. ResMed. AirView-Support your patients from diagnosis through ongoing care 2024 [Available from: https://www.resmed.com/en-us/healthcare-professional/products-and-support/monitoring-and-data-management/airview/.
35. Schwab RJ, Badr SM, Epstein LJ, Gay PC, Gozal D, Kohler M, et al. An official American Thoracic Society statement: continuous positive airway pressure adherence tracking systems. The optimal monitoring strategies and outcome measures in adults. Am J Respir Crit Care Med. 2013;188(5):613-20.
36. Selim B, Hilbert J, Yaggi HK. Principles of Noninvasive Pressure-targeted Ventilation. In: Grippi MA, Antin-Ozerkis DE, Dela Cruz CS, Kotloff RM, Kotton CN, Pack AI, editors. Fishman's Pulmonary Diseases and Disorders, 6e. New York, NY: McGraw-Hill Education; 2023.
37. Ramirez A, Khirani S, Aloui S, Delord V, Borel JC, Pépin JL, et al. Continuous positive airway pressure and noninvasive ventilation adherence in children. Sleep medicine. 2013;14(12):1290-4.
38. Bedi PK, DeHaan K, Ofosu D, Olmstead D, MacLean JE, Castro-Codesal M. Predictors of NIV-related adverse events in children using long-term noninvasive ventilation. Pediatric pulmonology. 2023;58(12):3549-59.
39. Atag E, Krivec U, Ersu R. Non-invasive Ventilation for Children With Chronic Lung Disease. Frontiers in Pediatrics. 2020;8.
40. Alberta TGo. $2M to kick-start new Edmonton children's hospital 2021 [updated July 29, 2021. Available from: https://www.alberta.ca/release.cfm?xID=796327B10F666-B9C2-1296-3BFFE56BF603137F.
41. Carmody JK, Duraccio KM, Krietsch KN, Simmons DM, Byars KC. Factors affecting pediatric adherence to positive airway pressure: Patient- and caregiver-reported treatment barriers and sleep difficulties. Sleep medicine. 2023;101:58-65.
42. Amin R, Al-Saleh S, Narang I. Domiciliary noninvasive positive airway pressure therapy in children. Pediatric pulmonology. 2016;51(4):335-48.
43. Hovenier R, Goto L, Huysmans T, van Gestel M, Klein-Blommert R, Markhorst D, et al. Reduced Air Leakage During Non-Invasive Ventilation Using a Simple Anesthetic Mask With 3D-Printed Adaptor in an Anthropometric Based Pediatric Head–Lung Model. Frontiers in Pediatrics. 2022;10.
44. Respironics. Interpreatation guide for Encore software compliance reports. 2014.
45. Ennis J, Rohde K, Chaput JP, Buchholz A, Katz SL. Facilitators and Barriers to Noninvasive Ventilation Adherence in Youth with Nocturnal Hypoventilation Secondary to Obesity or Neuromuscular Disease. Journal of clinical sleep medicine : JCSM : official publication of the American Academy of Sleep Medicine. 2015;11(12):1409-16.
46. Pascoe JE, Sawnani H, Hater B, Sketch M, Modi AC. Understanding adherence to noninvasive ventilation in youth with Duchenne muscular dystrophy. Pediatric pulmonology. 2019;54(12):2035-43.
47. Nicki B, Matt W, Heather E, editors. A Review of the Benefits, Challenges and the Future for Interfaces for Long Term Non-Invasive Ventilation in Children2018.
48. IBM Corp. IBM SPSS Statistics for Windows, Version 29.0.2.0. Armonk, NY: IBM Corp; 2023.

49. Cammarota G, Simonte R, De Robertis E. Comfort During Non-invasive Ventilation. Front Med (Lausanne). 2022;9:874250.
50. MacLean JE, Fauroux B. Long-term non-invasive ventilation in children: Transition from hospital to home. Paediatric respiratory reviews. 2023;47:3-10.
51. Amaddeo A, Khirani S, Griffon L, Teng T, Lanzeray A, Fauroux B. Non-invasive Ventilation and CPAP Failure in Children and Indications for Invasive Ventilation. Front Pediatr. 2020;8:544921.
52. Muller GJ, Hovenier R, Spijker J, van Gestel M, Klein-Blommert R, Markhorst D, et al. Non-invasive Ventilation for Pediatric Hypoxic Acute Respiratory Failure Using a Simple Anesthetic Mask With 3D Printed Adaptor: A Case Report. Front Pediatr. 2021;9:710829.
53. Laverty A, Gannon J, Robinson V, Samuels M, Abel F, Chan E. Adherence to long term non-invasive ventilation in children. European Respiratory Journal. 2015;46:PA1564.
54. Vincent M, Davies P, Gibson N, Morley A. G379 Descriptive analysis of adherence with non-invasive ventilation in children. Archives of Disease in Childhood. 2015;100(Suppl 3):A154-A5.
55. Simon SL, Duncan CL, Janicke DM, Wagner MH. Barriers to treatment of paediatric obstructive sleep apnoea: Development of the adherence barriers to continuous positive airway pressure (CPAP) questionnaire. Sleep medicine. 2012;13(2):172-7.
56. Marcus CL, Radcliffe J, Konstantinopoulou S, Beck SE, Cornaglia MA, Traylor J, et al. Effects of positive airway pressure therapy on neurobehavioral outcomes in children with obstructive sleep apnea. Am J Respir Crit Care Med. 2012;185(9):998-1003.
57. Fauroux B, Cutrera R. Editorial: Pediatric Long-Term Non-invasive Ventilation. Front Pediatr. 2021;9:654578.
58. Guilleminault C, Pelayo R, Clerk A, Leger D, Bocian RC. Home nasal continuous positive airway pressure in infants with sleep-disordered breathing. J Pediatr. 1995;127(6):905-12.
59. Markström A, Sundell K, Stenberg N, Katz-Salamon M. Long-term non-invasive positive airway pressure ventilation in infants. Acta Paediatr. 2008;97(12):1658-62.
60. Mc Namara F, Sullivan CE. Obstructive Sleep Apnea in Infants and Its Management With Nasal Continuous Positive Airway Pressure. Chest. 1999;116(1):10-6.
61. Hill NS. Chapter 18. Noninvasive Positive-Pressure Ventilation. In: Tobin MJ, editor. Principles and Practice of Mechanical Ventilation, 3e. New York, NY: The McGraw-Hill Companies; 2013.
62. Pinto VL, Sharma S. Continuous Positive Airway Pressure. StatPearls. Treasure Island (FL): StatPearls Publishing
Copyright © 2024, StatPearls Publishing LLC.; 2024.
63. Ramirez A, Delord V, Khirani S, Leroux K, Cassier S, Kadlub N, et al. Interfaces for long-term noninvasive positive pressure ventilation in children. Intensive Care Medicine. 2012;38(4):655-62.

Appendix A: Glossary of Terms

The following NIV terms used throughout the thesis are commonly used in literature, and they are defined as follows:

(1) **Non-invasive ventilation (NIV)** refers to the provision of mechanical ventilation without requiring an invasive artificial airway. Non-invasive positive-pressure ventilation using a mask (or *interface*) that conducts room air from a positive-pressure ventilator into the nose or mouth has become the predominant means of administering NIV worldwide (7). NIV has become an integral component of breathing support in acute and chronic settings because it avoids the complications of invasive ventilation (61). For this thesis proposal, NIV will refer to an umbrella of therapies, including continuous positive airway pressure (CPAP) and bilevel positive airway pressure (BIPAP).

Continuous positive airway pressure (CPAP) is a type of positive airway pressure used to deliver a set pressure to the airways maintained during inspiration (16, 62). Applying CPAP maintains positive end-expiratory pressure; it can decrease atelectasis, increase the alveolus's surface area, improve ventilation-perfusion matching, and improve oxygenation (16, 62).

(2) **Bilevel Positive Airway Pressure (BIPAP)** is pressure-limited ventilation. BIPAP delivers a cycling, pre-set pressure support which is higher during inspiration than expiration (16, 42). The high airflow provided by the ventilator augments the patient's inspiratory efforts. It should be delivered synchronously with the patient's respiratory efforts (16, 42). BIPAP is generally indicated when chronic hypercapnic respiratory failures occur from various etiologies, and CPAP is not tolerated or ameliorates upper airway obstruction (42).

(3) **Headgear** refers to the straps/headpiece used to keep the NIV mask in place. Headgear is designed to minimize discomfort and optimize the placement and stabilization of the NIV mask (9, 11).

(4) **Masks interface** refers to the equipment resting on the infant or child's face through which positive airway pressure is delivered (9, 11, 22, 63). Four standard mask interface types are available for home NIV use in children, including nasal masks, oro-nasal masks, nasal pillows, and total-faced masks (9, 11, 16, 63).

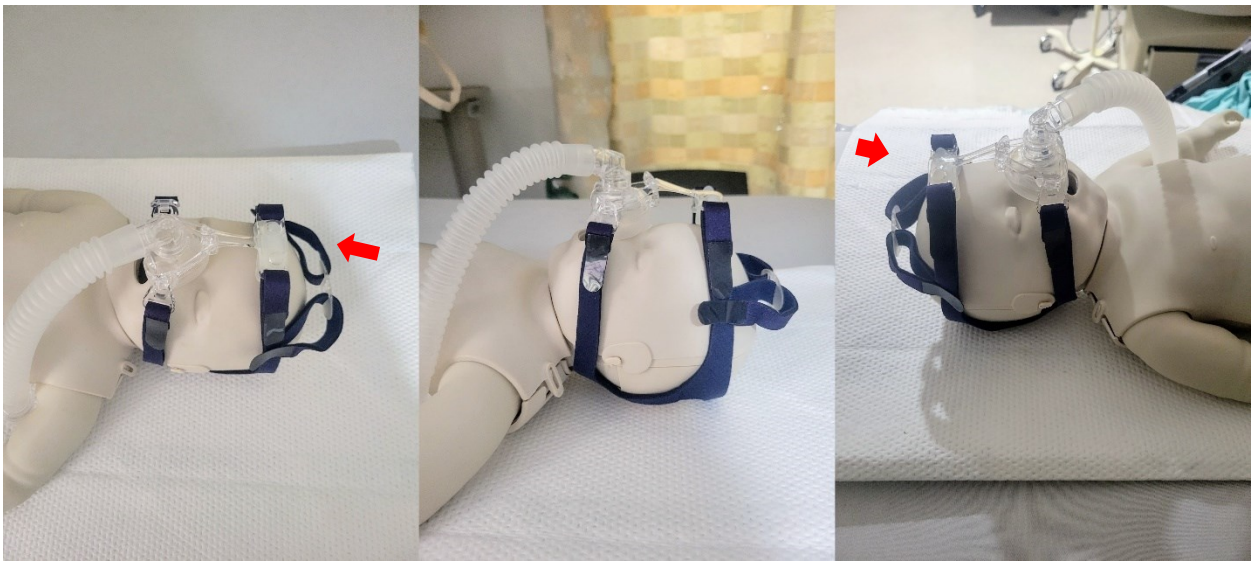
(5) **Adherence** refers to NIV therapy usage by infants and children in this thesis. Definitions of adherence vary in the literature. For this thesis, acceptable adherence is using NIV for at least four hours a night on 70% of nights (6, 18, 37).

Appendix B: Examples of Currently Available NIV Mask Fit and Headgear Adaptation on Mannequins

Figure 3. Poor Infant NIV Mask Fit and Headgear Adaptation #1.

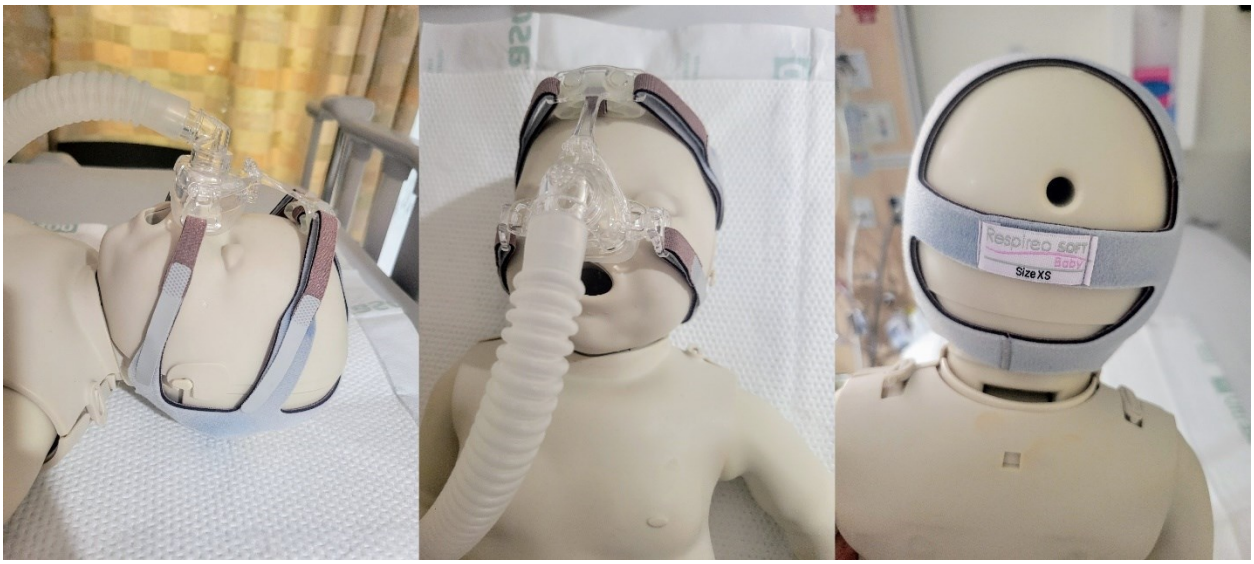
Note: This infant mannequin was wearing a small Respireo nasal cushion and a small Respireo headgear as a set. The nasal cushion fit the infant; however, the headgear was too large, as indicated by the red arrows, which led to mask displacement.

Figure 4. Poor Infant NIV Mask Fit and Headgear Adaptation #2.



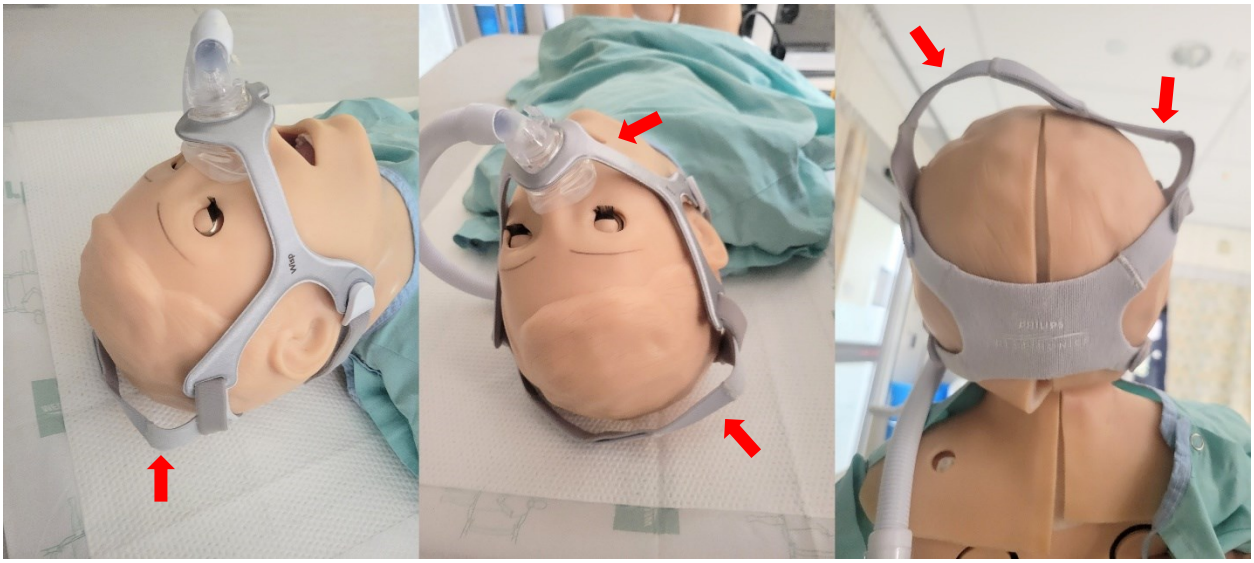
Note: This infant mannequin was wearing a small Cirri nasal cushion and a small Cirri headgear as a set. The nasal cushion fit the infant; however, the headgear was too large, as indicated by the red arrows, which made this set unusable.

Figure 5. Good Infant NIV Mask Fit and Headgear Adaptation.



Note: After having the Stollery NIV program fit the mask and headgear, it was determined that this infant mannequin needed a small Cirri nasal cushion with an extra small Respireo headgear to obtain a proper nasal cushion and good headgear fit as indicated.

Figure 6. Poor Child NIV Mask Fit and Headgear Adaptation #1.

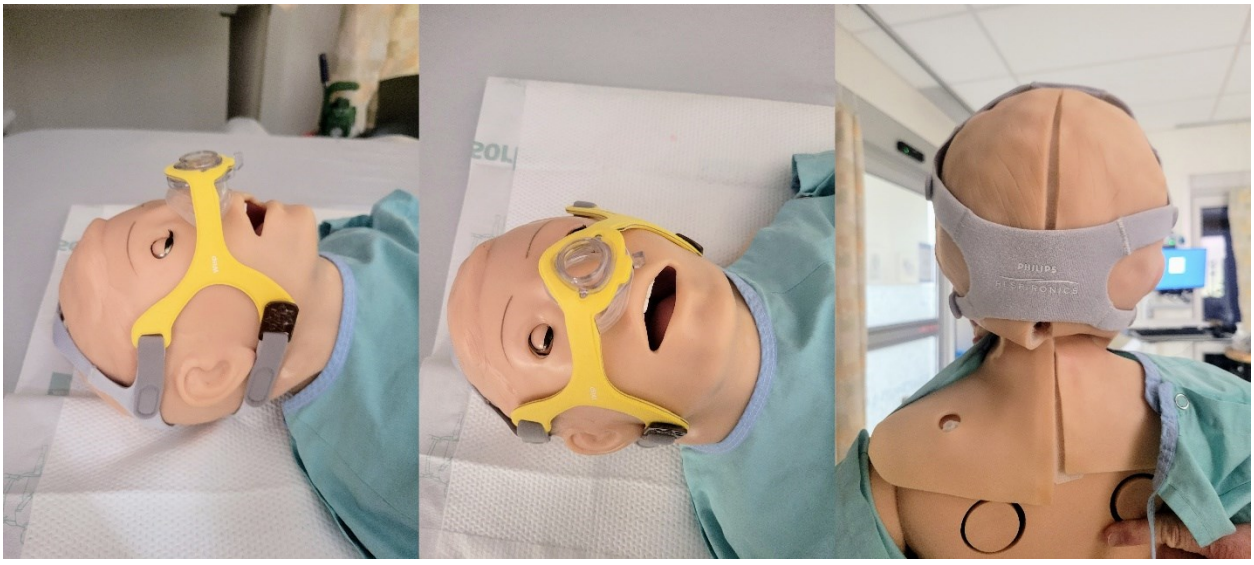


Note: This child mannequin was wearing a youth Wisp nasal mask and a youth Wisp headgear as a set. The nasal cushion fit the child; however, the frame and the headgear were too large, as indicated by the red arrows, which made this set unusable.

Figure 7. Poor Child NIV Mask Fit and Headgear Adaptation #2.

Note: This child mannequin was wearing a pediatric Wisp nasal mask and a pediatric headgear as a set. The nasal cushion fit the child; however, the frame and the headgear were too small, as indicated by the red arrows, making this set inappropriate for the child.

Figure 8. Good Child NIV Mask Fit and Headgear Adaptation.



Note: After having the Stollery NIV program fit the mask and headgear, it was determined that this child mannequin needed a pediatric Wisp nasal mask with a youth Wisp headgear to obtain a proper nasal cushion and good headgear fit as indicated.

Appendix C: REDCap Forms with Collected Variables

Demographics

Record ID	_____
Last Name	_____
First Name	_____
Date of Birth	_____
Alberta Healthcare Number	_____
Sex	○ Male ○ Female
Geographic Location	○ Edmonton ○ Within Alberta ○ Out of Province
Primary Reason for NIV Initiation	_____

NIV Therapy

Record ID	_____
Initiation Date	_____
Date of Maskfit	_____
Additional Maskfit	_____
Additional Maskfit 2	_____
Age at NIV Initiation	○ < 2 years ○ 2 years to < 5 years ○ 5 years to < 13 years ○ 13 years and up
Initiation Location	○ Inpatient ○ Outpatient ○ Not Documented
Therapy Type	○ CPAP ○ BPAP
Mask Type	○ nasal ○ full face
Headgear Adaptation needed?	○ Yes ○ No
Case or Control Group	○ Case ○ Control
Did patient use NIV therapy based on clinical documentation?	○ Yes ○ No
Did patient use NIV therapy within the first 1 month after NIV recommendation?	○ Yes ○ No
Did the patient use NIV therapy within the first 3 months after the NIV recommendation?	○ Yes ○ No
Did the patient use NIV therapy within the first 6 months after the NIV recommendation?	○ Yes ○ No

Reason for No NIV start	◯ Improved condition ◯ Inpatient use only ◯ NIV initiated before 2012 or after 2015 ◯ Transferred care ◯ Deceased or need more therapy ◯ Lost to follow up
Reason for No NIV adherence downloads	◯ With other sleep vendors ◯ Missing data due to system upgrades

Visits and Adherence

Record ID	_____
1 month adherence download available?	○ Yes ○ No
Download Date	_____
Days Included	_____
Total Average Hours of Use	_____
Days used	_____
Average Hours of Use Days Used	_____
Percentage of Nights with Use >4 Hours	_____
3 months adherence download available?	○ Yes ○ No
Download Date	_____
Days Included	_____
Total Average Hours of Use	_____
Days used	_____
Average Hours of Use Days Used	_____
Percentage of Nights with Use >4 Hours	_____
6 months adherence download available?	○ Yes ○ No
Download Date	_____

Days Included	_____
Total Average Hours of Use	_____
Days used	_____
Average Hours of Use Days Used	_____
Percentage of Nights with Use >4 Hours	_____
Discontinued within 6 months of starting	○ Yes ○ No