PRACTICE GUIDELINES FOR THE CARE OF PATIENTS UNDERGOING MANDIBULAR DISTRACTION OSTEOGENESIS

A DOCTORAL PROJECT

Submitted in Partial Fulfillment of the Requirements

For the degree of

DOCTOR OF NURSING PRACTICE

By

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ABSTRACT

Background

The purpose of this project was to develop evidence-based multidisciplinary clinical practice guidelines to direct the post-operative care of children with Robin sequence (RS) who undergo mandibular distraction osteogenesis (MDO). The care of the patient with RS who undergoes MDO is a very complex process, and one that requires multiple disciplines and pediatric subspecialists. The goal of this project was to use evidence-based strategies and expert opinion (where evidence was not available) to improve the clinical care of this complex population of children who have RS. The Iowa Model provided the necessary structure to develop the RS practice guidelines.

Methods

This project took place at a private, non-profit tertiary children’s hospital located in an urban area of Los Angeles. This hospital subscribes to the concept of family-centered care. Children with RS requiring MDO receive care in the newborn and infant critical care unit (NICCU) and pediatric intensive care unit (PICU). The guidelines were developed for children with RS younger than 6 months with airway obstruction caused by micrognathia. Approximately twelve to fifteen patients with RS undergo MDO yearly at this institution. The stakeholders for this project included a multidisciplinary team of providers and the families of patients undergoing MDO.
Results

The successful post-operative management of complex patients requires clear guidelines. In developing practice guidelines that mitigate variability in care, it was essential to involve the entire multidisciplinary team. Nursing has an opportunity to participate in the development of multidisciplinary practice guidelines to insure comprehensive care. The resulting clinical practice guidelines are comprehensive and inclusive of all stakeholders. Stakeholders have approved the proposed guidelines and they are now ready for piloting.

Discussion/Conclusion

It is projected that utilizing evidence-based interdisciplinary clinical practice guidelines to direct the post-operative care of children with RS who undergo MDO will significantly improve the quality and care coordination of these complex children. The next steps will be to pilot the guidelines on two to three infants. Feedback from all of the key stakeholders will be critical during this stage, as having their support will help ensure that these proposed guidelines become the new standard of care for the RS population. Once in place, an ongoing evaluation of this new standard of practice will be part of the plan, as well as the dissemination of our results on a national and international level.
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Embarking on this DNP journey has been life changing for me and would not have been possible without the support and guidance of many people. I would like to thank Dr. Kathy Hinoki, my chair for her continuous support throughout this project. Her attention to detail and gentle guidance helped to make this project a reality. I would also like to thank Dr. Jean O’Neil for her insightful comments and encouragement. To my fellow doctoral students for their collaboration, support, and friendship, you made this journey fun. Many thanks to Children’s Hospital Los Angeles, Craniofacial and Cleft Center and the team of providers who participated in this project. To my family and friends who encouraged me when the journey became difficult.

A heartfelt thank you to my parents Tom and Helen Clarke who believed in me and always encouraged me to follow my dreams. A very special thank you to Larry, Megan and Ellen, for providing me with unfailing love, support and continuous encouragement, this accomplishment would not have been possible without you. Thank you.
BACKGROUND

Robin sequence (RS) is defined as the presence of micrognathia (small-receded mandible), glossoptosis (posterior placement of the tongue) and airway obstruction (Breugem et al., 2016). Many children diagnosed with Robin sequence also present with a U-shaped cleft palate. All neonates with RS require assessment and management of airway obstruction. Mandibular Distraction Osteogenesis (MDO) is a surgical intervention for airway obstruction in infants born with Robin sequence that is used when interventions that are more conservative are unsuccessful (Tahiri, Greathouse, & Tholpady, 2015). Breugem et al. (2016) estimate that the prevalence of RS is about 1 in 8,500 live births.

The main goal of MDO is to increase the airway at the tongue-base by gradually increasing the length of the mandible (Hammoudeh et al., 2012). MDO is a surgical technique that involves a mandibular osteotomy and placement of distraction devices, followed by a short period of healing. After one to two days, activation of the distractors begins, and the osteotomy is opened 1.8 mm each day, during which time the tongue moves forward and the airway opens. A consolidation phase of 12 weeks follows, when ossification of the mandible occurs. The benefits of using MDO to treat airway obstruction are described in the surgical literature (Tahiri et al., 2015); however, postoperative management practices following this procedure have not yet been established.
**Problem Statement**

The MDO surgery post-operative management is the responsibility of the plastic surgery/craniofacial team, led by the craniofacial fellow who has typically completed a plastic surgery residency and who is seeking an additional year of subspecialty training in craniofacial surgery. The post-operative course is highly dependent upon the surgical specialists and the management plan that is determined by the team. No standardized post-operative plan of care currently exists for this complex patient population. The recovery period is often fragmented and prolonged. It is additionally unclear whether the post-operative clinical staff has a complete understanding of the procedure or of the potential post-operative complications.

Post-operative care of the RS population is a very complex process, and one that requires several disciplines and pediatric subspecialists. It is believed that the provision of standardized care for infants who undergo MDO surgery will likely improve patient outcomes and contribute to family centered care. The typical inpatient stay of six weeks has the potential of being reduced, an outcome that could result in a significant reduction in hospital costs and resources.

**Purpose Statement**

The purpose of this DNP project was to develop a set of standardized practice guidelines for the post-surgical care of patients undergoing Mandibular Distraction Osteogenesis (MDO) and prepare it for implementation. The goals of developing a practice guideline were to mitigate variability in care, shorten the hospital length of stay, decrease overall hospital costs, and improve the quality of care. Meetings with the main stakeholders (maxillofacial surgeons, craniofacial fellows, pulmonologists,
neonatologists, otolaryngologist, craniofacial pediatrician, occupational therapist, nurse care managers, nurse practitioners, clinical care coordinators, craniofacial nutritionist and nursing administrators) were conducted to present the practice guideline, gain consensus from the team and plan its implementation. It was projected that going through this process would result in a more cohesive multidisciplinary team and the establishment of best practice standards for infants with RS, using an evidence-based approach.

**Supporting Framework**

The theoretical model selected to guide this project was The Iowa Model (Figure 1). This model has previously been used for the development of clinical practice guidelines in the literature and is familiar to many nursing groups (Bonnel & Smith, 2014). The Iowa Model was first developed in the early 1990s by nurse clinicians at the University of Iowa Hospitals and Clinics. It has been thought of as a pragmatic method for employing Evidence-Based Practice (EBP) to solve clinical problems, based upon current research. This model was originally published in 2001, and then updated as the Iowa Model Revised: Evidence-Based Practice to Promote Excellence in Patient Care in 2015. This specific model leveraged the unique contributions of the multidisciplinary team approach during the process of developing the clinical practice guidelines.

The Iowa model consists of seven steps (Doody & Doody, 2011). Step one was choosing an issue or problem that would be addressed using an EBP approach. The problem this project addressed was the fragmented care of patient undergoing MDO. The providers caring for this patient population agreed that improvements to standardize the post-operative management would be beneficial. Step two was the formation of a team. The RS team included a multidisciplinary team of stakeholders, including plastic/
maxillofacial surgeons, a pulmonologist, neonatologists, otolaryngologist, craniofacial pediatricians, craniofacial nurses, nutritionist, occupational therapist, clinical care coordinators, nursing administrators and staff from the newborn and infant critical care unit (NICCU). The study institution’s neonatal intensive care unit (NICU) is referred to as NICCU and is the unit where most of these patients are treated. Step three involved exploration and synthesis of the body of evidence including research, established guidelines and clinical standards. Step four evaluated whether the quality and strength of the evidence was enough to continue with the project.

Step five included the design and development of a plan to pilot the clinical practice guideline. After an extensive review of the literature, guidelines were developed with input from the team, based upon the best evidence available. When evidence was not available, expert opinion was utilized. This critical step considered the preferences of all stakeholders, including the family, as the EBP approach included being family centered. Provider and parent education, and the development of the implementation and evaluation plans occurred during this step. Step six would involve actual implementation of the clinical care guidelines. Support from all stakeholders would be critical during this stage, as having these new guidelines become the standard of care would be essential to ensure sustainability. This step would also include addressing barriers to implementation. Finally, step seven would involve ongoing evaluation of this new standard of practice and dissemination of the results. While the goal of this doctoral project was to complete the Iowa Model through step five, it is the intention of this doctoral student to complete the entire process post-graduation.
Figure 1. Iowa Model Revised.

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REVIEW OF LITERATURE

A systematic review of the literature was conducted to develop practice guidelines for the post-operative care of children undergoing mandibular distraction osteogenesis. The author searched the following databases: PubMed, Scopus, CINAHL and Google Scholar. Search terms included Pierre Robin Sequence, Robin sequence, mandibular distraction osteogenesis, osteogenesis, micrognathia, jaw abnormalities and airway obstruction. Additional search terms included surgical management, nursing management, patient outcomes, impact on the family, family centered care and post-operative course. This search was limited to peer reviewed, English language journals, publications between 2000 and 2017, and a population of children whose ages were limited from birth-23 months. Reference lists from pertinent articles were then reviewed to identify other relevant publications. The following topics were used to organize the review of the literature: background, definition of Robin Sequence, medical issues, medical and surgical management, family centered care and multidisciplinary care.

Overview

The cause or pathogenetic event resulting in RS is unknown. The generally accepted explanation is that prior to the tenth gestational week, micrognathia causes the tongue to be placed upward and posterior, preventing the palatine shelves from closing (Evans et al., 2011). This theory explains that the tongue physically preventing the palatal shelves from closing causes the U-shaped cleft, often seen in patients with RS. Molecular genetic studies are needed to understand the pathogenesis of RS (Evans et al., 2011).
**Definition of Robin Sequence**

The definition of RS is controversial. There is limited consensus regarding diagnostic criteria or evaluation of children with RS (Breugem et al., 2016; Izumi et al., 2012). The main point of disagreement is whether the patient must have a cleft palate to have the diagnosis of RS (Mackay, 2011). The lack of agreement about what constitutes the diagnosis itself makes it difficult to know the true incidence of RS. Estimates range from 1 in 8,500 to 1 in 20,000 births (Glenn et al., 2011; Evans et al., 2011; Breugem et al., 2016). An international, multidisciplinary clinical consensus group met in the Netherlands in 2014 to develop a clinical consensus report (CCR) and to identify best practices in the care of infants with RS. The CCR was based upon a synthesis of the literature and use of expert opinion (Breugem et al., 2016). The consensus group came to an agreement that RS would be diagnosed in children who are born with micrognathia (small, receded mandible), glossoptosis (tongue blocking posterior pharyngeal space), and upper airway obstruction. Cleft palate is also frequently seen, but is not necessary to make the diagnosis of RS.

**Medical Issues**

**Airway Issues**

The two main medical issues for infants with RS are upper airway obstruction and feeding challenges. If left unaddressed, they can lead to serious consequences including asphyxia, hypoxia, respiratory failure, cor pulmonale, failure to thrive and death (Evans et al., 2011). The assessment for airway obstruction must be a priority. Unfortunately, there are no widely accepted protocols to assess the airway in infants with RS.
Polysomnography (PSG) is used at many hospitals to diagnose airway obstruction and other sleep disorders, including central apnea in infants.

Because of limited access to PSG, there is a lack of universal acceptance that this diagnostic tool is necessary to diagnose airway compromise in infants with RS (Breugem et al., 2016). Glynn et al. (2011) noted that they did not have access to PSG and suspect that marginal airways may not be identified by physical examination alone. Anderson et al. (2011) studied a group of infants with RS who underwent PSG prior to airway management and found a high incidence of obstructive sleep apnea. Their findings suggest that obstructive sleep apnea (OSA) may be more common in children with RS than was previously thought. They also noted that snoring was not a good predictor of OSA in this population, as 45% of the patients studied had OSA without snoring. Many patients with RS have undiagnosed OSA and these findings support the importance of polysomnographic screening to identify OSA in patients with RS.

**Feeding Issues**

Feeding challenges in infants with RS are well described in the literature (Anderson et al., 2011; Breugem et al., 2016; Evans et al, 2011; Filip et al., 2015; Glenn et al., 2011; Mackay, 2011) and are mainly attributed to airway obstruction. Feeding issues include an inability to consume adequate nutrition by mouth for normal growth and development. Feeding problems include prolonged feeding times, insufficient oral intake, and unsafe oral feedings which can lead to respiratory distress. Some affected children are at risk for aspiration secondary to uncoordinated suck-swallow-breath during feedings or from a swallowing dysfunction (Breugem et al., 2016; Evans et al, 2011). It is estimated that 38% to 70% of infants with RS have feeding issues severe enough to warrant tube feeding.
(Evans et al., 2011; Filip et al., 2015). Gastro-esophageal reflux disorder (GERD) is also more common in infants with RS (Break, Umapathysivam, Tivey, & Anderson, 2016) and may further complicate feeding. Early feeding evaluation of infants with RS is essential. Infants who are identified as being unsafe to orally feed should be tube fed until it is determined to be safe to take fluids by mouth.

**Medical Management of Robin Sequence**

Medical management of airway obstruction in patients with RS includes non-surgical measures such as prone or side positioning, and the use of a nasopharyngeal airway. Patients who continue to have airway obstruction are considered for surgical procedures, in order to avoid a tracheostomy. Surgical procedures include tongue lip adhesion, subperiosteal release of the floor of the mouth, mandibular distraction osteogenesis (MDO) and tracheostomy (Anderson et al., 2011; Almajed et al., 2017).

A second medical issue for infants with RS involves feeding challenges and resulting issues of poor growth. Feeding issues improved after MDO, 82 percent of patients were exclusively oral feeding after surgery (Breik et al., 2016). Patients with isolated RS demonstrated greater improvement than those with genetic syndromes. Commonly there is a post-operative decline in growth during the first six weeks, warranting close follow up after undergoing MDO repair, especially during the first 6-8 weeks of the recovery period. Almajed et al (2017) & Breik et al (2016) reported improvement in airway, feeding and GERD symptoms after MDO. Although feeding challenges improve after MDO, close monitoring of growth and development is warranted.
Surgical Management of Robin Sequence

The effectiveness of the use of MDO to treat micrognathia in RS has been studied extensively. Systematic reviews by Almajed et al. (2017); Breik et al. (2016); and Tsui et al. (2016) found that MDO was effective in treating children with upper airway obstruction that was not responsive to conservative management. Early surgery was safe, effective and prevented the need for tracheostomy in 95% of the cases. Children who had associated genetic syndromes had a higher failure rate as compared to children with isolated RS (Hammoudeh et al., 2012). Failure was attributed to previously undiagnosed lower airway obstruction, central apnea, cardiovascular co-morbidities, and previously undiagnosed neurologic abnormalities (Breik et al., 2016). Cognitive delays have been reported in patients with RS. It is unclear if these delays were caused by repeated hypoxia or from an underlying developmental issue (Almajed et al., 2017).

Family Centered Care

Family-centered care (FCC) is described as a method to provide health care that respects, and is responsive to, the needs and values of families (Davidson, et al., 2017). It involves a partnership for health care planning and decision making (Kuo, et al., 2012) based upon the belief that the family is the constant in the child’s life and are the child’s source of support (Neft, Eichner, Hardy, & Klein, 2003). Davidson, et al. (2017) developed guidelines for family-centered care in the NICU with the goal to identify best practices for family-centered care in the NICU based on existing evidence. The guidelines support providing family members of infants in the NICU the choice of learning how to help provide care to their child. Parental presence and involvement in their child’s care in the NICU has been shown to improve the parent’s confidence and
ability to provide care to their child and has been shown to improve the mental health of parents during and after the hospitalization. The guidelines also support including family education programs as part of clinical care in the NICU as they have been shown to be helpful for families by decreasing depression, anxiety, stress and post-traumatic stress and increasing satisfaction with care (Davidson, et al., 2017; Bastani, Abadi & Bastani, 2015). Davidson, et al. (2017) reported that the impact of educational programs on length of stay (LOS) and cost is not clear and requires further study.

**Multidisciplinary Care of the Child with Robin Sequence**

Many of the articles reviewed promoted multidisciplinary care when treating the child with RS. However, there was a dearth of knowledge regarding the role of the nurse on the team. One article by Marcellus (2001) discussed implications for nursing practice, but it was written prior to the use of MDO as a treatment for these patients. No other articles discussed the role of the nurse in the care of the patient with RS. The parent and family perspective of having a child with RS who underwent MDO was not commonly seen in the literature. Hong, McNeil, Kearns & Magit (2012) looked at quality of life (QOL) by measuring health-related QOL after MDO as reported by parents. These researchers reported a subjective overall benefit after MDO.

In summary, a review of the literature identified agreements, discrepancies, as well as knowledge gaps on the topic of medical management of the child with Robin sequence. What is most critical to note is that there is a lack of consensus as to what constitutes best practice for diagnosing, evaluating and treating infants born with RS. Publications by Breugem et al. (2016) and Evans et al. (2011) have proposed plans to evaluate and manage patients with RS, but both groups acknowledged the need for
prospective studies to guide the development of evidence-based guidelines for the care of children with RS. The role of nurses and advanced practice nurses in caring for this population needs further exploration, and the experiences of the family whose child undergoes MDO needs further evaluation as well.
METHODS

The purpose of this project was to develop evidence-based clinical practice guidelines to direct the post-operative care of children with RS who undergo MDO. The goal of this project was to use evidence-based strategies and expert opinion (where evidence was not available) to improve the clinical care of this complex population of children who have RS. This section includes information about the population of children served and the hospital where the surgery and post-operative management took place. The Iowa Model provided the necessary structure to develop the RS practice guidelines as described in the methodology procedure section.

Setting

This DNP project took place at a private, non-profit tertiary children’s hospital located in an urban area of Los Angeles. This hospital subscribes to the concept of family centered care. More than 70% of patients served are Medicaid funded and over half of the families do not speak English at home. In addition to multiple outpatient clinics, children with RS receive care in the newborn and infant critical care unit (NICCU), pediatric intensive care unit (PICU) and a variety of the medical and surgical floors. Approximately twelve patients with RS undergo MDO yearly at this institution.

Stakeholders

The stakeholders for this project included the multidisciplinary team of providers and the families of patients undergoing MDO. The multidisciplinary team included medical and surgical specialists from maxillofacial/plastic surgery, otolaryngology, pulmonary/sleeps medicine, neonatology, pediatrics, occupational therapists, nutritionist, advanced practice nurses, staff nurses, nurse managers and family members. All
stakeholders contributed to the guidelines aimed at facilitating the care of these complex children. Parents of current patients undergoing MDO were interviewed and their input was incorporated into the guidelines.

**Population of Interest**

Since the objective of this DNP project was to develop practice guidelines for a specific set of children born with RS, there was no actual study sample involved. However, the practice guideline was developed for these particular children based upon established criteria. The inclusion criteria were as follows:

- Children born with RS, younger than 6 months
- Require MDO to manage airway obstruction caused by micrognathia
- Absence of anatomical airway anomalies (upper or lower)
- Gastro esophageal reflux disease (GERD) has been treated with H2 blocker
- Stable from a cardiac perspective
- No weight or age requirement
- Tracheostomy (will not preclude MDO)
- CT indicates: good bone stock, hypoplasia of the mandible. Decreased airway space

The exclusion criteria included:

- Major central nervous system (CNS) findings or intra cranial anomalies (with poor prognosis as determined by genetics or neurology)
- CT indicates bony defects
- Combination of CNS and laryngomalacia
- Significant central sleep apnea and or lower anatomical airway anomalies
Ethical Considerations

Applications were submitted to the medical facility's Institutional Review Board (IRB), in addition to the IRB at California State University, Los Angeles (CSULA) for their respective reviews. Since the purpose of this DNP project was to develop evidence-based practice guidelines for the RS population, the primary IRB institution made the decision that this project did not constitute research and was granted exempt status. The CSULA IRB found that this DNP project did not involve confidentiality or anonymity issues, nor did it contain any patient, family, or staff data. They were therefore in agreement with the primary IRB facility's decision of exemption.

Procedures

The Iowa Model provided the framework for developing the practice guidelines for infants with RS who require MDO, in a step-like fashion. While there are seven steps in the Iowa model, this project had as its goal the completion of only the first five parts. Step one involved choosing a problem to be addressed through EBP. The problem this project aimed to improve was the fragmented care of patients undergoing MDO. Step two was to form a team. The team involved interested stakeholders who agreed to aid with the development of practice guidelines. The multidisciplinary teams consisted of providers from plastic/maxillofacial surgery, pulmonary/sleep medicine, pediatrics, neonatology, otolaryngology, nutrition, occupational therapy, craniofacial advanced practice nurses (APN), nurse managers, NICCU clinical care coordinator and staff nurses who care for these patients.

Step three included exploring, evaluating and synthesizing the body of evidence including research, established guidelines and clinical standards. The review of the
literature was expanded as evidence was published/discovered. Step four evaluated whether the quality and strength of the evidence from the literature review was sufficient to continue with the project. The guidelines were developed upon the best available evidence at the time and utilized the preferences and expert opinion of the key stakeholders. Quality of the evidence was evaluated using the American Association of Critical Care Nurses (AACN) New Evidence Leveling System as seen in Table 1 (Armola et al., 2009).

Table 1

AACN’s New Evidence-Leveling System (Armola et al., 2009)

<table>
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<tr>
<th>Level</th>
<th>Description</th>
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<tr>
<td>Level A</td>
<td>Meta-analysis of multiple controlled studies or meta-syntheses of qualitative studies with results that consistently support a specific action, intervention or treatment</td>
</tr>
<tr>
<td>Level B</td>
<td>Well designed controlled studies, both randomized and nonrandomized, with results that consistently support a specific action, intervention, or treatment</td>
</tr>
<tr>
<td>Level C</td>
<td>Qualitative studies, descriptive or correlational studied, integrative reviews, systematic reviews, or randomized controlled trials with inconsistent results</td>
</tr>
<tr>
<td>Level D</td>
<td>Peer-reviewed professional organizational standards, with clinical studies to support recommendations</td>
</tr>
<tr>
<td>Level E</td>
<td>Theory-based evidence from expert opinion or multiple case reports</td>
</tr>
<tr>
<td>Level M</td>
<td>Manufacturers’ recommendations only</td>
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</table>
Step five involved the design of a family centered clinical practice guideline and provider and parent educational materials. Implementation and evaluation plans were also developed during this step. In order to accommodate the busy schedule of the key stakeholders, the DNP student met with each person individually to obtain pertinent feedback in a timely manner. This project used a consensus model to identify critical components of the post-operative care and to elicit agreement from stakeholders. To achieve this goal, the DNP student followed these steps:

- Conducted individual interviews with key stakeholders (plastic and maxillofacial surgeons, otolaryngologist, pediatric subspecialists, craniofacial PNP's, craniofacial nutritionist, occupational therapist, NICCU nursing providers and nurse managers) to determine successful practices and areas needing improvement.
- Drafted practice guidelines based upon published evidence and key stakeholders’ expert opinions and preferences.
- Evaluated the quality of the evidence using the American Association of Critical Care Nurses (AACN) New Evidence Leveling System (Armola et al., 2009).
- Presented the first iteration of the guidelines, to the craniofacial fellow and the plastic surgeon in order to make needed edits and to obtain their endorsement.
- Edited the document based upon input from each stakeholder.
- Circulated a word copy of the guidelines to key stakeholders for comment.
- Continued to revise the guidelines until a consensus was gained.
- Sought agreement from key stakeholders regarding timelines for implementation.
- Developed educational materials targeting providers and staff.
- Developed education materials for families, incorporating these new guidelines.
RESULTS

This DNP project was conducted through step five of the Iowa model, which was the goal of this DNP project. The first four steps went smoothly. Step five the design and development of the clinical practice guideline and provider and parent educational materials took much longer than anticipated secondary to delays in meeting with individual key stakeholders. The resulting products are comprehensive and inclusive of multiple disciplines and providers. This DNP project will be deemed successful if the RS practice guidelines have been established, approved by the major stakeholders for implementation.

During the development of these guidelines, an unintended consequence occurred. Optimal pain management during the distraction process was identified as a topic of concern. Some stakeholders believed that the infants were receiving too much medication during the distraction process, leading to prolonged sedation and delayed oral feeding. The NICCU staff will address optimal pain assessment and management separately. Another observation was the frequent delay in the surgical removal of the distraction devices by the three months recommended timeline. This was due to multiple factors, including patients being lost to follow up and delayed surgery-scheduling secondary to patient illness. A quality improvement project will take place to mediate this variance.
IMPLEMENTATION PLAN

Step six will involve the pilot and actual implementation of the clinical care guidelines. Support from all stakeholders will be critical during this stage, as having these new guidelines become the standard of care will be essential to ensure sustainability. This step will also include addressing barriers to implementation. Finally, step seven will involve ongoing evaluation of this new standard of practice and dissemination of the results. While the goal of this doctoral project was to complete the Iowa Model through step five, it is the intention of this doctoral student to complete the entire process post-graduation.” The DNP student will follow the steps described below to complete the project:

• Provided education regarding MDO and the plan of care, as described in the guidelines, to the parent during a 1:1 education session. Materials written at the 4th grade reading level in English and Spanish incorporate photos and diagrams. Model, with the actual distractors, used during the educational session allow family members to handle the distractors on the model before being asked to care for their child with the distractors in place.

• Pilot approved guidelines with two subsequent patients admitted for MDO.

• Revise guidelines based upon input from stakeholders involved in the care of the two pilot patients.

• Edited educational materials and guidelines based upon feedback.

• Guidelines become the standard of care.

• Incorporate RS MDO guidelines into the EMR.
ANTICIPATED RESULTS

The standardized clinical care guidelines that were developed for the care of infants with RS who undergo MDO will likely improve patient outcomes and contribute to family centered care. The parent education provided will potentially decrease the anxiety that parents of infants undergoing this procedure experience. It is anticipated that the nurses caring for these babies will have a better understanding of the procedure and will be able to provide comfort to their patients without the use of excessive sedation. Early incorporation of the dysphagia team in feeding therapy will aid in the safe transition from enteral to oral feedings. Reduction of the current lengthy inpatient stay could result in a significant reduction in hospital costs and resources. The guidelines also included information about care of the infant with RS after MDO and discharge. Appointments for follow up care and surgical removal of the distraction device will be scheduled prior to discharge. This discharge planning will provide clear instructions for parents regarding the care of their child. During the admission, the parent will have met members of the Craniofacial and Cleft Center including advance practice nurses who will provide a link to the multidisciplinary team who will provide care for their child future needs.
METHOD FOR EVALUATING THE NEW PRACTICE GUIDELINES

When instituting changes in patient care practices, it is crucial to perform a formal evaluation to assess whether patient outcomes have improved, worsened, or remained the same. The timing of the evaluation phase of the project would be dependent upon the number of patients referred for MDO treatment, under these new practice guidelines. It is anticipated that within six months of having implemented the new RS guidelines, that at least two patients would have undergone MDO surgery. It is believed that this number of patients would make for an adequate trial. A comparison of patient outcomes based on pre-guidelines versus post implementation would then take place.

In regards to patient outcomes, hospital length of stay (LOS) of the pilot patients would be compared to patients treated prior to implementation of these guidelines. Additionally, parent satisfaction would be measured and compared, using the Consumer Assessment of Health Care Providers and Systems (CAHPS) Child Hospital Survey, the satisfaction tool employed at the hospital. Stakeholder interviews will provide feedback regarding their satisfaction and use of the guidelines. Modifications to the guidelines will be made prior to implementation. Lastly, when the practice guidelines gain acceptance into the medical facility's plan of care, the author would meet with information technology (IT) to plan for the integration of the RS practice guidelines tool into the institution's electronic medical record (EMR) system.
DISCUSSION/RECOMMENDATIONS

The successful post-operative management of complex patients requires clear guidelines. In developing practice guidelines that mitigate variability in care, it is essential to involve the entire multidisciplinary team. Nursing has an opportunity to participate in the development of multidisciplinary practice guidelines to insure comprehensive care. In an effort to deliver family centered care, the family must be involved in the development of practice guidelines as well.

The care of the patient with RS who undergoes MDO is a very complex process, and one that requires multiple disciplines and pediatric subspecialists. Once the guidelines are shown to be effective at the current institution, they will be offered to other centers performing this procedure. An abstract describing our experience developing and implementing the guidelines will be submitted, for consideration, as a podium presentation at the next Annual American Cleft Palate and Craniofacial Association (ACPA) meeting in 2019. When there is enough experience to show positive results, a manuscript will be written, and submitted for publication in The Cleft Palate-Craniofacial Journal.
REFERENCES


## APPENDIX A

**CARE GUIDELINES FOR INFANTS WITH ROBIN SEQUENCE (RS) REQUIRING MANDIBULAR DISTRACTION OSTEOGENESIS (MDO)**

### Patients:
- Usually <30 days of age
- 90% inpatient vs. 10% outpatient referral (+/- supplemental oxygen)
- **Respiratory Distress** (patient on oxygen) vs. **Respiratory Failure** (patient intubated)
- In some cases, the patient is situated prone or in lateral position in the NICCU before MDO surgery

### Pre-Operative Consultations (requested by NICCU) Before MDO
Respective specialist will order the following tests as needed

#### Plastic Surgery (PS):
- Interdisciplinary conference (within 4 days of admission, based on patient need) including Neonatology, Plastic Surgery, Craniofacial NP and Pulmonology
- Craniofacial NP will consult with family and provide education. Topics covered will include RS and MDO, introduction to Craniofacial Team Center care, and ongoing outpatient case management
- Craniofacial NP to consult with family prior to discharge

#### Pulmonology:
- Chest x-ray to rule out intrinsic lung disease
- Baseline sleep study:
  - Evaluate for central apnea, obstructive apnea, Apnea-Hypopnea Index (AHI)
  - Indicated if no ET tube and/or minimal oxygen
  - Not indicated if patient intubated for airway management

#### Otolaryngology:
- Request rigid L&B (scheduled prior to MDO)
- R/O other airway abnormalities
- Findings may change the decision of whether distraction is indicated

#### Radiology:
- Maxillofacial CT without contrast

#### Genetics:
- Identification of potential comorbidities

#### Cardiac Anesthesia:
- If cardiac anomalies are present, obtain cardiac anesthesia clearance

#### Dysphagia:
- Dysphagia evaluation
- If not intubated and safe to do so, begin therapy to address feeding
**Indications for Surgery**

**Mandibular Distraction Osteogenesis**

### Indications:
- Absence of anatomical airway anomalies (upper or lower)
- Gastroesophageal reflux disease (GERD) has been treated with H2 blocker
- Stable from a cardiac prospective
- No weight or age requirement
- Tracheostomy (will not preclude MDO)
- CT indicates: good bone stock, hypoplasia of the mandible, decreased airway space

### Not Indicated:
- Major central nervous (CNS) findings or intracranial anomalies (w/poor prognosis determined by genetics or neurology)
- Tracheostomy may be best treatment for these patients
- CT indicates bony defects
- Combination of CNS findings and laryngomalacia
- Significant central sleep apnea and or lower anatomical airway anomalies

### Surgery:
- Lasts about 3 hours
- Patient will go from ICU → OR → ICU (85% NICCU, about 15% PICU)
- Rigid L&B by Otolaryngology and intubation as indicated
- If no significant lower airway abnormalities, Plastic Surgery will proceed with osteotomy and placement of distractors
**Post-Operative Guidelines**

**Distraction Process:**

- **Latency Period ~ 48 hours**

Activation of distraction initiated by the PS fellow (or resident) and then daily (~0800) to assess the patient and proper functioning of the distractor. The PS will call the bedside RN with a time estimate of their arrival so that the patient may receive comfort measures prior to the distractor advancement.

Pain assessment and titrate for comfort per unit protocol:
- After 72 hours post-op, non-narcotic (i.e. Acetaminophen, sweet-ease) to be used as the primary agent for discomfort.

The distractor, subsequently activated every 8 hours by the beside RN or PS Fellow. It is turned two turns (0.3 mm/turn) three times a day (total 1.8 mm daily at 0800, 1600 and midnight).

Each time the distractor is turned (PS, RN or parent) provider will record it in the device diary.

- Patient will return to the OR (PS fellow to be present) to be extubated, usually 7-10 days after starting distraction

---

**Pin care:**
- Before every turn, the nurse will remove existing xeroform (preventing it from becoming stuck in the mechanism)
- Clean pin site with half strength H2O2 and apply new xeroform
- When distraction is completed, distraction arms will be removed by the PS fellow (may be done at bedside). If there is a possibility that the skin can be closed over the distractor, then removal and closure in OR

**Feeding (ordered by neonatologist):**
- Dysphagia evaluation (OT or SLP) the day after extubation, if airway is stable
- Start NG feeding PO day 1 if safe to do so
- Daily dysphagia therapy for feeding
Transfer from PS Service:
- Continue dysphagia therapy for feeding – NG or bottle
- Continue reflux therapy (H2 blocker)
- Airway
  - Schedule PSG if there is a question of residual obstructive sleep apnea (OSA) or need for oxygen (O2)
    - May go home on low flow O2 w/pulse oximeter secondary to risk for central apnea with desaturation even after distraction
    - If there is a question of residual OSA or need for O2, a PSG is needed prior to DC

Otherwise...
- PSG to be scheduled any time after:
  - Distraction is complete
  - Patient off sedation/narcotics
  - Preference when NG is out

Hospital Discharge:
- When airway and feedings are stable
- Prior to DC, the Clinical care Coordinator (CCC) will schedule the following appointments:

- PMD within 5 days for close monitoring of growth, development and GERD management (Provide PMD w/info sheet on MDO)
- PS, Craniofacial PNP & Nutritionist 2 weeks after DC
  - PS To fill out booking slip for surgery schedulers to schedule removal of distractors 3 months after consolidation (device removal may be scheduled w/o post-distraction sleep study. If post-distraction sleep study not done, patient will be scheduled for prolonged stay in PACU)

- Pulmonary (include nurse care manager (NCM)) 4-6 weeks after discharge
  - Schedule sleep study prior to cleft palate (CP) repair

- G-tube care after DC according to provider who placed it
  - G-tube peg → GI
  - G-tube → Pediatric Surgery

- Craniofacial Team Appointment
  - Schedule at 3-4 months of age (prior to removing distractor)
  - Team to include (as a minimum) evaluation by: PS, Pulmonary, PNP, SW, Nutrition
**Surgical Removal of Distraction Device:**
- Patients scheduled for device removal without a post-distraction sleep study will be scheduled for a prolonged PACU stay
- If patient is on O2, they will stay overnight
- No post-operative wound care
- Scar managed with massaging and taping
- Continue supportive care for feeding and development with PMD

**Cleft Palate Repair (if needed):**
- Post-distraction sleep study must take place before cleft palate repair
- Cleft Palate repair will be scheduled when patient is 12-15 months old and Pulmonologist/PSG indicated it safe to do so
APPENDIX B

*Parent Education: Infants with Robin Sequence (RS) Requiring Mandibular Distraction Osteogenesis (MDO)*

Robin Sequence is a condition where babies are born with a small lower jaw that is set back causing the tongue to be placed in the back of the mouth toward the throat causing the airway to become blocked. A cleft palate may also occur. RS occurs in 1 in 8,500 babies born.

Mandibular Distraction Osteogenesis (MDO) is a surgical procedure done to improve airway obstruction in infants born with Robin sequence. It is used when other measures do not work.

MDO works by gradually increasing the length of the lower jaw and guiding the tongue forward, which allows the size of the airway to increase.

- Surgery lasts about 3 hours.
- An incision is made under the jaw line, the jawbone is separated and a distraction device is attached to either side of the separated bone. This will take place on both sides of the jaw.
- The distraction device will not be seen after the surgery, two small rods called distractor arms, will come out behind the ear on each side.
- A short period of healing will follow when a callus forms.
- After about 48 hours, the distractor arms will begin to be turned.
- Distractor arms are turned, every eight hours and the space is opened 0.6mm each time (total of 1.8 mm per day). During the time between turns, the jawbone will grow to fill this space.
• As the jaw is lengthened the tongue moves forward and the airway opens
• If the child has a breathing tube, they will be monitored and the tube will be removed when it is safe to do so.
• When the jaw length is to the desired amount, the distraction arms will be removed.
• The internal distraction device will remain in place for 12 weeks, after turning has stopped, to allow the new bone to become strong.
APPENDIX C

NURSING EDUCATION: INFANTS WITH ROBIN SEQUENCE (RS) REQUIRING MANDIBULAR DISTRACTION OSTEOGENESIS (MDO)

Patients:
- Usually <30 days of age
- 90% inpatient vs. 10% outpatient referral (+/- supplemental oxygen)
- Respiratory Distress (patient on oxygen) vs. Respiratory Failure (patient intubated)
- In some cases, the patient is situated prone or in lateral position in the NICCU before MDO surgery

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Plastic Surgery (PS):
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- Craniofacial NP to consult with family prior to discharge to review outpatient care

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- Baseline sleep study:
  - Evaluate for central apnea, obstructive apnea, Apnea-Hypopnea Index (AHI)
  - Indicated if no ET tube and/or minimal oxygen
  - Sleep study not indicated if patient intubated for airway management

Otolaryngology:
- Request rigid L&B by Otolaryngology (scheduled just prior to MDO)
- Rule out other airway abnormalities, which could contribute/explain, airway obstruction.
- Findings may change the decision of whether distraction is indicated

Cardiac Anesthesia:
- If cardiac anomalies are present, obtain cardiac anesthesia clearance prior to surgery

Radiology:
- Maxillofacial CT without contrast

Genetics:
- Identification of potential comorbidities

Dysphagia:
- Dysphagia evaluation
- If not intubated and safe to do so, begin therapy to address feeding
**Indications for Surgery**

**Mandibular Distraction**

**Osteogenesis**

**Indications:**
- Absence of anatomical airway anomalies (upper or lower)
- Gastroesophageal reflux disease (GERD) has been treated with H2 blocker
- Stable from a cardiac prospective
- No weight or age requirement
- Tracheostomy (will not preclude MDO)
- CT indicates: good bone stock, hypoplasia of the mandible, decreased airway space

**Not Indicated:**
- Major central nervous (CNS) findings or intracranial anomalies (w/poor prognosis determined by genetics or neurology)
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**Surgery:**
- Lasts about 3 hours
- Patient will go from ICU → OR → ICU (85% NICCU, about 15% PICU)
- Rigid L&B by Otolaryngology and intubation as indicated
- If no significant lower airway abnormalities, Plastic Surgery will proceed with osteotomy and placement of distractors
**Mandibular Distraction Osteogenesis (MDO)**

**Post-Operative Guidelines**

**Distraction Process:**

- **Latency Period ~ 48 hours**

activation of distraction initiated by the PS fellow (or resident) and then daily (~0800) to assess the patient and proper functioning of the distractor. The PS will call the bedside RN with a time estimate of their arrival so that the patient may receive comfort measures prior to the distractor advancement.

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- Dysphagia evaluation (OT or SLP) the day after extubation, if airway is stable
- Start NG feeding PO day 1 if safe to do so
- Daily dysphagia therapy for feeding

**Hospital Discharge:**
- When airway and feedings are stable
- Prior to DC, the Clinical care Coordinator (CCC) will schedule the
**APPENDIX D**

**TABLE OF EVIDENCE**

<table>
<thead>
<tr>
<th>Purpose (author, date)</th>
<th>Method</th>
<th>Sample</th>
<th>Results</th>
<th>Conclusions/Limitations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Systematic review to determine the success of MDO in treating upper airway obstruction in children with micrognathia Breik (2016)</td>
<td>Search databases: CINAHL, PubMed, SCOPUS, EMBASE, Web of Knowledge and gray literature sources Scirus and Med Nar</td>
<td>PICO Criteria- Pt. inclusion: micrognathia, UAO, failed conservative tx., s/p b/l MDO, +/- syndromes, &amp; gt;1 year f/u</td>
<td>MDO prevented trach in 95% of cases</td>
<td>MDO very effective at preventing trach in children with UAO not responsive to conservative tx.</td>
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<td></td>
<td>Studies included were published and unpublished in English language, from 1990-2013</td>
<td>Exclusion: known pre-op central apnea or lower airway abnormalities, unilateral MDO, other jaw conditions causing UAO other than micrognathia.</td>
<td>Syndromic pt. 4x failure rate vs. isolated RS</td>
<td>Ideal age for MDO not established (&lt;18 months no difference in failure rate).</td>
</tr>
<tr>
<td></td>
<td>2 independent reviewers</td>
<td>Intervention: b/l MDO Comparator: trach</td>
<td>Failure caused by: previously un dx. lower airway obstruction, central apnea, cardiovascular co-morbidities, and undiagnosed neurologic abnormalities.</td>
<td>Early surgical correction appears to be safe.</td>
</tr>
<tr>
<td></td>
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<td>Outcomes: airway status, primary MDO, decanulation of trach</td>
<td>Not as successful in decannulating trach pt. (81% success rate) secondary to pre-op GERD, issues with swallowing and trach complications.</td>
<td>In trach group &lt;24 mo. Were most successfully decannulated.</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Types of studies: RCT, quasi-experimental, prospective and retrospective cohort, and case control</td>
<td>&gt;24 months less successful</td>
<td>Limitations: None discussed</td>
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<tr>
<td></td>
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<td>66 studies included</td>
<td></td>
<td>Recommendations: Need for long term studies were discussed.</td>
</tr>
<tr>
<td>Purpose (author, date)</td>
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<td>Results</td>
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<tr>
<td>Systematic review to determine the success of MDO in treating feeding problems and GERD in children with micrognathia Breik, (2016)</td>
<td>Search databases: CINAHL, PubMed, SCOPUS, EMBASE, Web of Knowledge and gray literature sources Scirus and Med Nar</td>
<td>This review is a subset of the larger systematic review by Breik et al (2016). Of the studies in the larger review, only those that included outcomes related to Feeding or GERD were included. Search terms: Feeding, gastroesophageal reflux, MDO, RS, syndromic micrognathia. Types of studies: case series and case reports. 21 studies included</td>
<td>MDO improves feeding (82% exclusive oral feeding post-surgery). Pt. with isolated RS fed better than those with syndromes (94% vs. 73%). Post op decline in growth during first 6 weeks. Improvement in GERD noted in 66/70 pt. post MDO</td>
<td>Improved airway function improves feeding and GERD. Findings provide only a guideline to factors associated with ↑ chance of failure and identify groups of pts. at ↑ risk for problems. Limitations: poor quality studies (case report &amp; case series). Incomplete data. Heterogeneity between RS &amp; pt. with syndromes grouping difficult. Recommendations: close nutrition and growth monitoring with feeding support after MDO especially the first 6-8 weeks after MDO. Further research is needed to understand the long-term success in improved feeding and resolution of GERD.</td>
</tr>
<tr>
<td>Purpose (author, date)</td>
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<tr>
<td>Systematic review to understand airway outcomes of surgeries addressing UAO in patients with RS. PO UAO measured by PSG Almahed, (2017)</td>
<td>Search databases: MEDLINE (PubMed interface) and Cochrane Library database. Authors also hand searched references. Studies included were published and unpublished in any language, from 1981-2015 2 independent reviewers</td>
<td>Inclusion criteria: studies reporting outcome of surgical treatment for mandibular hypoplasia in RS. Exclusion: case reports, surgery &gt;12 months Search terms: Pierre Robin syndrome, distraction osteogenesis, tongue, tongue lip adhesion, subperiosteal release of the floor of the mouth Types of studies: cohort studies or case series 70 studies were included (46 on MDO)</td>
<td>Mortality and morbidity (trach) were low in all surgical procedures. MDO had lowest issues with PO airway obstruction (3.6%)</td>
<td>Pt with RS have persistent central apnea through the 1st year of life. Cognitive delays have been reported in pt. with RS. It is unclear if caused by repeated hypoxia or an underlying developmental issue. Limitations: no RCT comparing procedures. All data was collected retrospectively. Lack of comprehensive data on PO treatment of UAO in RS using PSG. Recommendations: Future studies to report PSG data: AHI, accurate documentation of PO airway obstruction. Future studies to report incidence of central apnea with AHI.</td>
</tr>
<tr>
<td>Purpose (author, date)</td>
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<tr>
<td>Systematic review to determine the Effectiveness of DO to treat OSA Tsui (2016)</td>
<td>Search databases: PubMed, Ovid, Cochrane Library, and Scopus. Systematic reviews were per PRISMA statement with 3 rounds of search &amp; evaluation the 1 round of critical appraisal. Also, used manual reviews. Studies were included prior to 8/12/2014. Did not restrict language, publication date, or status of publication. 2 independent reviewers</td>
<td>Inclusion: human studies, clear description of tx., pre-op and PO AHI reported, f/u duration was reported. Exclusion inclusion criteria Search terms: distraction, distraction osteogenesis, sleep apnea, airway obstruction Type of studies: clinical trials &amp; case series on tx. and outcome of OSA with MDO. 12 studies were included</td>
<td>All studies reported 90-100% success of MDO in OSA pt. Complication rate was 0-21% mostly from wound infection and numbness. All studies used PSG to dx OSA. Mean pre-op AHI 10-50/h ↓ to 1.1-5/h after MDO Endoscopy used to assess airway dimension and any abnormal pathology. Different distraction protocols were included.</td>
<td>MDO showed hopeful results in children tx with OSA. Limitations: wide age ranged from 7 days to 60 years. Criteria defining severity of OSA, tx success/failure was not defined for infants &amp; children. Recommendations: Need RCT to compare MSO with traditional maxillomandibular advancement surgery.</td>
</tr>
</tbody>
</table>
### Table 2

**Retrospective reviews: Treatment of Upper Airway Obstruction with Mandibular Distraction Osteogenesis**

<table>
<thead>
<tr>
<th>Purpose/ Resource</th>
<th>Design/Key Variables</th>
<th>Sample/Setting</th>
<th>Measures</th>
<th>Findings</th>
<th>Conclusions/Study Limitations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Retrospective review to determine effectiveness of internal mandibular distractors in treatment of infants with OSA</td>
<td>Retrospective review IV: OSA symptoms DV: internal mandibular distraction</td>
<td>29 infants with OSA treated at CHLA (Los Angeles) between 2003 &amp; 2007. 19 had RS and 10 had a variety of other syndromes/health issues All received multidisciplinary evaluation prior to MDO. Pulmonary/critical care, otolaryngology, Plastics/maxillofacial Surgery.</td>
<td>PSG on all patients. AHI determined by # of OSA, mixed apnea and hypopneas per hour. Detailed distraction protocol included.</td>
<td>All pt. AHI improved after MDO. Mean pre-op AHI in respiratory distress group-39.7 &amp; PO AHI-5.8). Mean Po AHI in respiratory failure group-3.13. 1patient went home on oxygen. LOS-42.3 days (range 3-140 days).</td>
<td>MDO relieved OSA in patients studied. Multidisciplinary approach is associated with lower complications. Limitations: retrospective review. Recommendations: Develop selection criteria for pt. prior to MDO.</td>
</tr>
<tr>
<td>Retrospective review to determine effectiveness and safety of MDO in infants with RS weighing &lt; 4 kg</td>
<td>Retrospective review IV: pt. wt. &lt;4 kg and &gt; 4 kg DV: complications and comorbidities</td>
<td>Infants with RS tx. with MDO at Riley Children’s Hospital (Indiana) between 2004 &amp; 2015. 81 weighed &lt; 4kg. 40 weighed &gt; 4kg (control) All received multidisciplinary evaluation prior to MDO including Pt. who failed conservative measures had PSG. Pt with no central apneas, AHI &gt; 20 or significant CO2 retention were considered for MDO. Maxillofacial CT with reconstruction and direct L&amp;B to r/o other causes of OSA.</td>
<td></td>
<td>All pt. AHI improved after MDO. No significant difference in success of MDO in pt. &lt; 4kg. compared to control.</td>
<td>MDO is safe and effective tx of OSA in infant’s w RS &lt; 4 kg. at experienced high volume centers with NICU and peds. Anesthesia Limitations: retrospective design, no long-term follow up. It is unknown</td>
</tr>
<tr>
<td>Purpose/ Resource</td>
<td>Design/Key Variables</td>
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<tr>
<td>To identify predictors of success and complications of MDO when treating micrognathia: -MDO before other airway procedure -Trach then MDO (Lam et al., 2014)</td>
<td>Retrospective cohort review</td>
<td>Infants with RS tx. with MDO at Cincinnati Children’s Hosp. between 1995 &amp; 2009. 61 in MDO 1st 62 in trach 1st group</td>
<td>Potential predictors of outcomes: demographic (age at time of MDO, sex), f/u time, syndrome (isolated RS &amp; other), initial surgery (MDO vs. trach), length of mandible distraction, # of distractions, endoscopic airway procedures, base of tongue procedures, repair of choanal atresia repair). Surgical success was defined as avoiding trach in MDO 1st group and decanulation in trach before MDO group</td>
<td>Trach 1st more likely to have a syndrome vs MDO group (66% vs. 43%). MDO group younger (5 mo. vs. 30 mo.) and requires fewer subsequent airway procedures. Overall complication rate 26.8% ↑in trach 1st group (38.7% vs. 14.8%)</td>
<td>MDO can avoid trach in many pts. Pts with syndromes and other co-morbidities required trach more often, were older at time of MDO, had more complications and ↓rate of decannulation. Limitations: retrospective design</td>
</tr>
<tr>
<td></td>
<td>Detailed distraction protocol included.</td>
<td></td>
<td>Complications were the same for both groups and included surgical site infection. Mean pre-op AHI in pt &lt; 4kg 41.5, vs.26.1 in control. PO AHI in &lt; 4kg. 12.1 vs. 12.4 in control</td>
<td>how MDO may affect future jaw growth. Recommendations: Close collaboration with anesthesia and NICU teams.</td>
<td></td>
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</tbody>
</table>
### Table 3

**Outcomes of Multidisciplinary Care of the Child with RS**

<table>
<thead>
<tr>
<th>Purpose/Resource</th>
<th>Design/Key Variables</th>
<th>Sample/Setting</th>
<th>Measures</th>
<th>Findings</th>
<th>Conclusions/Study</th>
</tr>
</thead>
<tbody>
<tr>
<td>To describe pt with RS and CP over a 33 year period from a multidisciplinary perspective (Filip et al., 2015)</td>
<td>Retrospective descriptive study</td>
<td>104 consecutive pts. with RS and CP born between 1980 &amp; 2010 treated at CL&amp;P unit Oslo, Norway. Multidisciplinary study team: PS, psychology, orthodontics, SLP and nursing.</td>
<td>Retrospective data collected and compared to a large control group for each specific variable.</td>
<td>Airway interventions (oropharyngeal or nasopharyngeal tube, CPAP, trach) used at the same rate for syndromic and non-syndromic pts. ↑incidence of feeding difficulties in RS like other studies. Feeding/growth-wt. at birth and at CP repair ↓ than the normative population. No growth spurt in 1st yr. of life, despite a ↓ in feeding difficulties. Strong association between boys with RS and ASD with 17.9% with ASD vs. 2.3% in boys with CP only.</td>
<td>Authors question if their respiratory and feeding interventions were adequate. PSG not standard of care for pt with RS. Standardized f/u for pt with RS regarding feeding and growth. Limitations: Retrospective design, missing or lost data Recommendations: Prospective trials needed to determine when respiratory interventions should be used. Investigate further the finding of ↑ASD in RS.</td>
</tr>
<tr>
<td>To examine multidisciplinary</td>
<td>Retrospective review</td>
<td>69 pts. with RS treated between 1991 &amp; 2010 at Children’s</td>
<td>Retrospective review of protocol to assess and</td>
<td>Airway management: 39% failed position tx, of those 59% were</td>
<td>Authors report that 84% of pt with RS were successfully treated</td>
</tr>
</tbody>
</table>
### Topics: Airway management, nutritional status, and hearing

Univ. Hospital Temple Street Dublin, Ireland

62 had isolated RS with CP, 4 had RS as part of a syndrome.

Multidisciplinary management of airway, feeding, and serous otitis media
Assessment & protocol included.

Multidisciplinary study team: PS, ENT, pediatrician, anesthesiologist, audiologist, SLP, nurse specialist.

manage 3 variables: the airway, nutrition hearing

Successfully tx. with nasopharyngeal airway. Of the remainder: 2 had glossopexy, 1 nasal continuous positive airway pressure, 12% trach.

Feeding: 70% NG/ G-tube, 30% special bottle,
35% of those successfully treated by positioning required supplemental feeding.

Audio: 35% needed PE tubes

To determine the cause of RS, in pt followed at 2 children’s hospitals, compare findings to reports in the literature, contrast interventions to pt with isolate vs. syndromic RS

(Izumi et al, 2012)

Retrospective chart review and systematic review of publishes articles on RS

Topics: Airway and feeding management

Comprehensive literature search was done to compare their finding to those reported in the literature.

Medical records of pt with RS from 2 children’s hospitals 28 pt- RBCH, Cleveland, Oh and 97 pt- RCH, San Diego, CA

Treatment of airway and feeding problems: prone position, MDO, trach, MDO& trach, tongue lip adhesion, NPA, ET intubation, tube feeding, death at birth

Positioning successful in 62% (RBCH-50%, RCH-65%).

Tube feeding required in 52% (RBCH-79%, RCH-43%).

Review of records at 2 sites as well as systematic literature review showed similar rates of each syndrome, about 40% had isolated RS, and 60% had

Importance of collecting family history and prenatal exposures during eval of RS to identify possible syndrome cases.

Pt with RS require aggressive tx of airway and feeding challenges.

Need for aggressive airway mgt and NG
additional syndromic features.

feeding more common in syndromic pts.
Mgt = institutional preference

Limitations:
retrospective reviews at tertiary hospitals may have more severe cases of RS.
No agreement of definition/EBP for care of child with RS.
No specific definition of micrognathia

Recommendations:
All pt with RS need eval by clinical geneticist

Note: AHI= Apnea-Hypopnea Index, ASD=autism spectrum disorder, b/l=bilateral, CHLA: Children’s Hospital Los Angeles, eval=evaluation, f/u=follow up, mgt= management, MDO= mandibular Distraction osteogenesis, mo.=month, NG= nasogastric tube, NP= nasopharyngeal, OSA= Obstructive Sleep Apnea, op=operative, pt=patient, peds=pediatrics, PS= plastic surgery, PO= post-operative, , PSG= polysomnography, PE tube= pressure equalizing tube, s/p=status post, RCH= Rady Children’s Hospital, RBCH= Rainbow Babies and Children’s Hospital, resp= respiratory, RS= Robin Sequence, tx= treatment, SPL= speech language pathologist, trach= tracheostomy, UAO=upper airway obstruction, vs.=versus.