



Children's Hospital Colorado

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BACKGROUND

Pierre Robin Sequence (PRS) refers to the triad of micrognathia, glossoptosis and airway obstruction. PRS commonly presents with a cleft palate or other craniofacial syndromes. PRS in neonates can lead to airway obstruction, poor feeding and poor growth. Conservative measures such as prone positioning, nasal trumpet, or high flow oxygen are often trialed, but upwards of 71% of patients may require surgical intervention.¹ Mandibular distraction osteogenesis (MDO) has emerged as an effective surgical alternative to tracheostomy and tongue-lip adhesion.

MDO is a process in which the mandible is surgically lengthened with surgical plates placed along the body of the mandible after controlled osteotomies. The mandible is then advanced anteriorly by separating the plates through rotating distractor pins. Advancement of the mandible pulls the tongue anteriorly, improving airway obstruction. Studies demonstrating the success of MDO in PRS neonates are limited by small sample sizes and there is debate over the effectiveness of MDO in improving airway, feeding and growth measures.^{2,3,4}

The Children's Hospital of Colorado Pediatric Otolaryngology Department was an early adopter of MDO and has a large cohort of patients who have undergone this treatment. The purpose of this investigation is to examine outcomes in neonates undergoing early (<90 days of life) MDO. This project is an extension of an initial reported cohort of 24 patients.⁵

METHODS

This study is a retrospective review over an 15-year period of patients with micrognathia who underwent MDO within the first 90 days of life (N=63).

Data Collected: Demographic data, hospital course variables, associated syndromes, duration of distraction, need for additional procedures, and growth curve data were included. The Cormack-Lehane classification was used to evaluate the grade of laryngeal view before and after MDO.

Statistics: Descriptive statistics along with mixed-effects growth curve analyses were utilized. Comparisons of estimated mean weights from the linear mixed effects regression model are compared to CDC norms using 2-sided 1 sample t-tests.

Study conducted under **COMIRB protocol #: 18-2660**

RESULTS

Table 1. Patient demographics, hospital stay and post-op outcomes in isolated and syndromic PRS subgroups

Covariate	Overall (N=63)	PRS-Isolated (N=50)	PRS-Syndromic (N=13)
Male	28 (44.4%)	22 (44.0%)	6 (46.2%)
Birth Weight (Kg)	2.80 (0.67)	2.80 (0.67)	2.77 (0.67)
Term (Weeks at Gestation)	37.9 (2.06)	38.2 (1.99)	36.9 (2.03)
Age at Surgery (Days)	36.0 (24.3)	34.4 (24.7)	41.9 (23.1)
Weight at Surgery (Kg)	3.47 (0.66)	3.45 (0.69)	3.51 (0.53)
Length of Distraction (Days)	7.62 (2.11)	7.76 (2.1)	7.08 (2.14)
Distance of Distraction (mm)	14.2 (3.0)	14.4 (3.0)	13.2 (2.6)
Post-op Day of Discharge	19.4 (11.9)	19.2 (11.9)	20.1 (12.1)
Oxygen at Discharge	4 (6.3%)	2 (4.0%)	2 (15.3%)
Post-op G-Tube required	13 (20.6%)	10 (20%)	3 (23.1%)
MDO Revision/Device Failure	7 (11.1%)	6 (12%)	1 (7.7%)
Repeat Distraction	0 (0%)	0 (0%)	2 (15.4%)

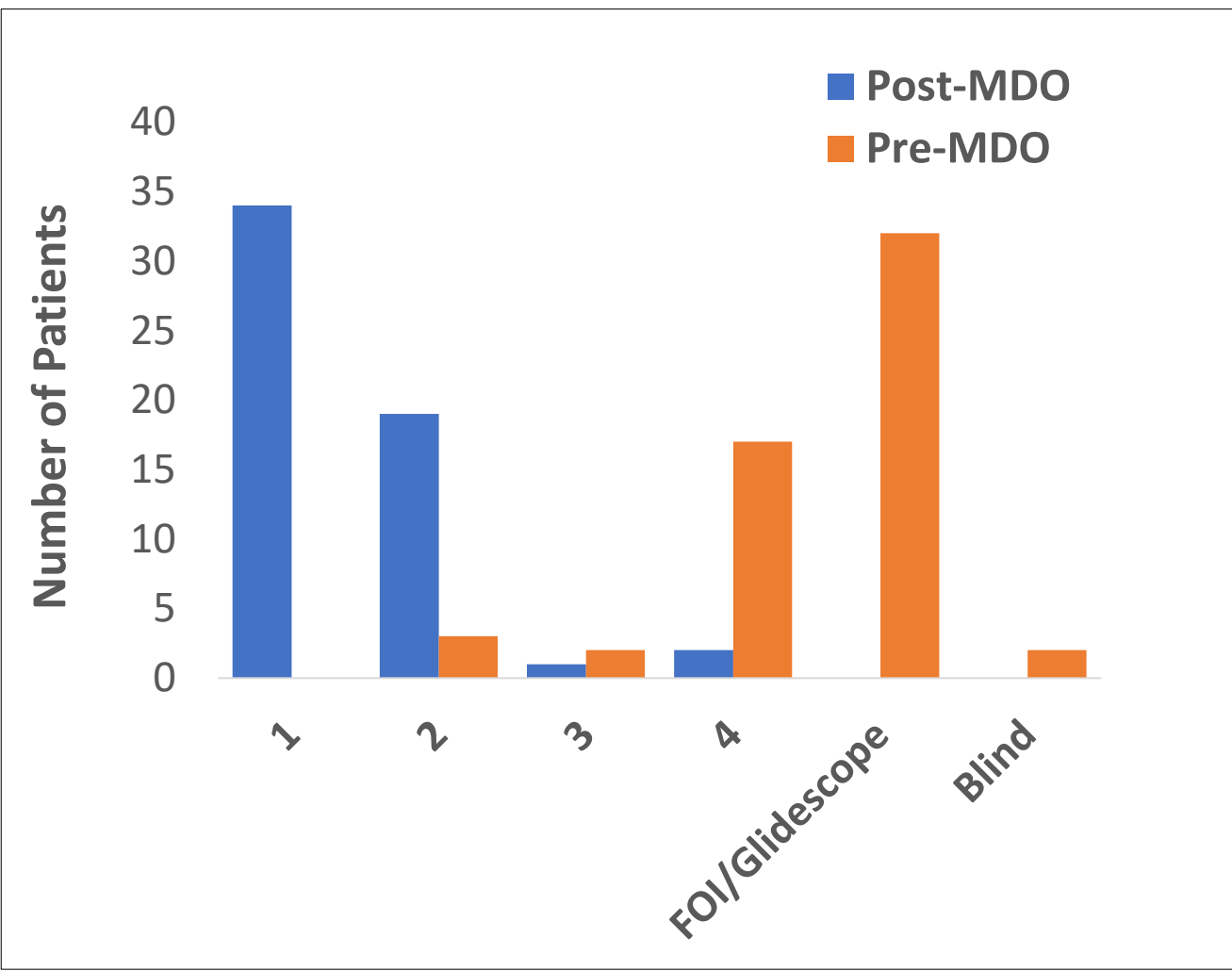


Figure 1. Airway grade by Cormack-Lehane classification before and after MDO. Grades 1 and 2 were considered “easy” and grades 3-4 and those only able to be intubated fiberoptically or with a glidescope were determined “difficult.”

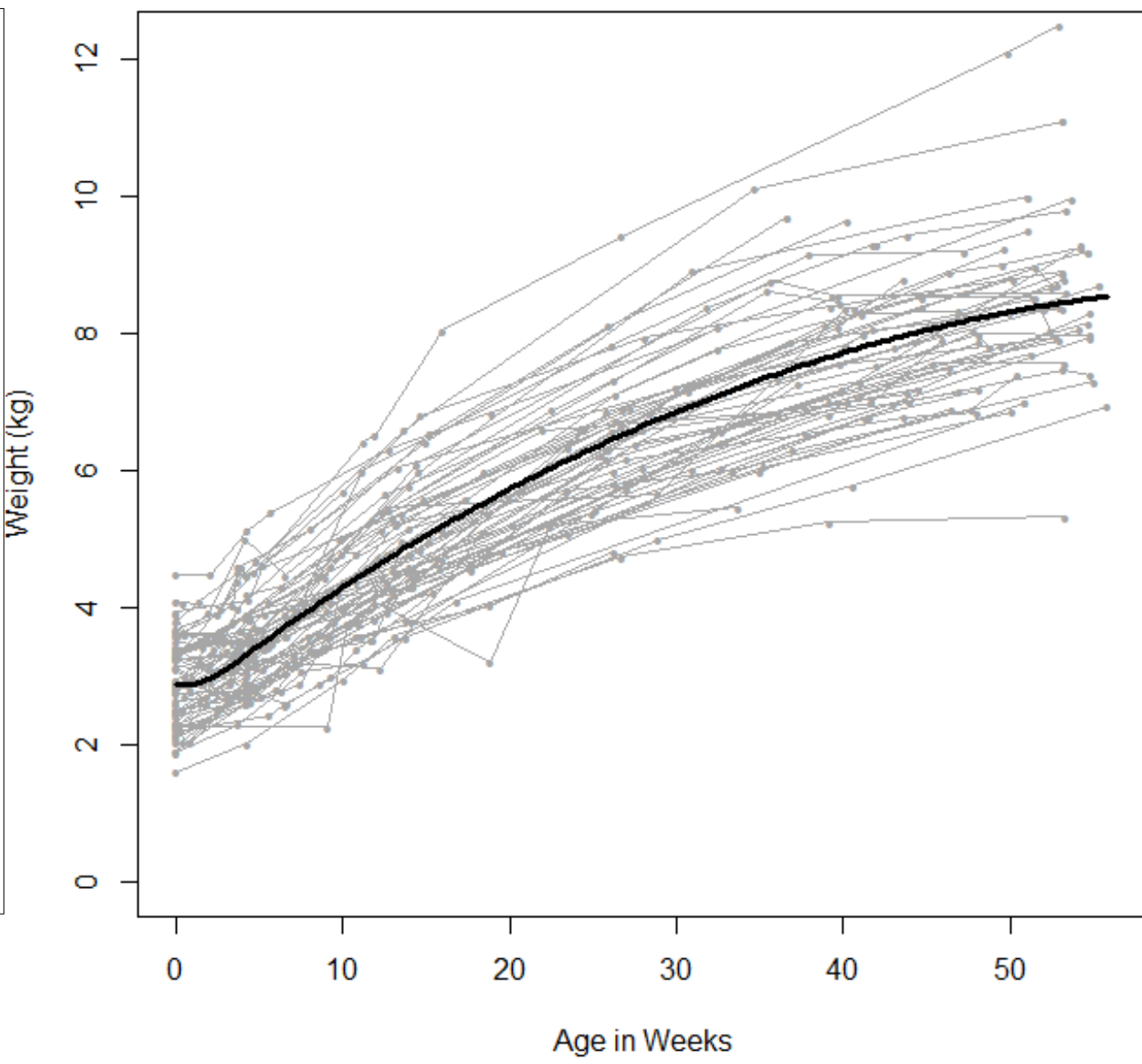


Figure 2. All patients' weight for age (N=63)

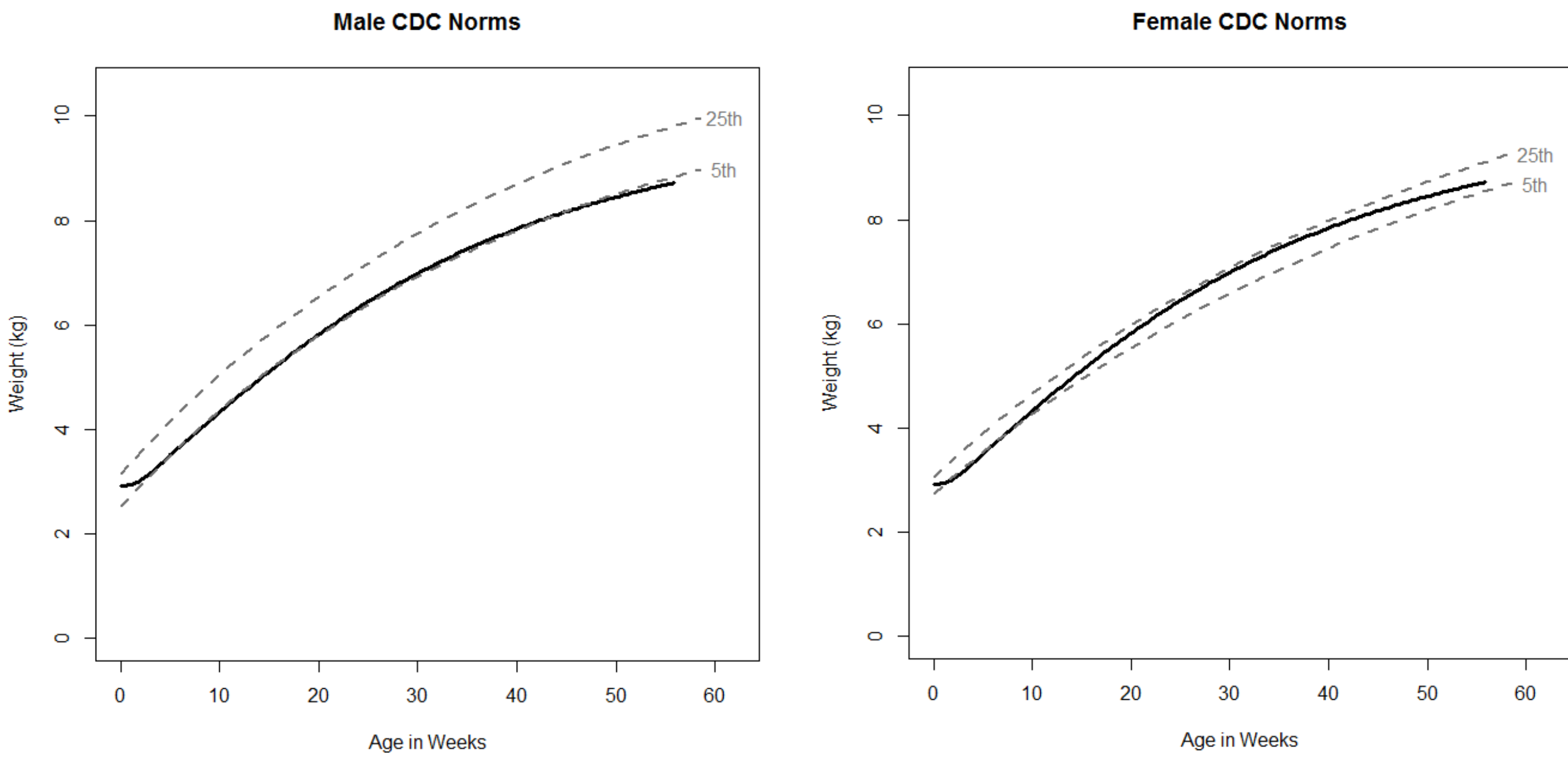


Figure 3 and 4. Isolated PRS patient' weight for age compared to male (n=22) and female (n=28) norms

Table 1. Isolated PRS estimated growth with CDC norm comparisons (n=50)

Time	Estimated Mean Weight in kg (95% Bootstrap CI)	Comparison to CDC Norm Weights in kg (p-value)			
		Male 5th Percentile	Male 25th Percentile	Female 10th Percentile	Female 25th Percentile
Birth	2.92 (2.68, 3.17)	2.36 (<0.001)	3.15 (0.079)	2.75 (0.166)	3.06 (0.270)
1 mo	3.38 (3.14, 3.61)	3.21 (0.145)	4.01 (<0.001)	3.44 (0.653)	3.79 (0.001)
3 mo	4.80 (4.56, 5.04)	4.67 (0.276)	5.53 (<0.001)	4.67 (0.290)	5.08 (0.027)
6 mo	6.56 (6.30, 6.81)	6.33 (0.088)	7.30 (<0.001)	6.19 (0.007)	6.66 (0.441)
9 mo	7.76 (7.52, 8.01)	7.53 (0.068)	8.60 (<0.001)	7.37 (0.003)	7.89 (0.323)
12 mo	8.55 (8.29, 8.79)	8.41 (0.287)	9.58 (<0.001)	8.30 (0.063)	8.86 (0.019)

CONCLUSIONS

These results support early MDO as a successful option for neonates with symptomatic micrognathia refractory to conservative measures. Improvement in airway grade as assessed independently by anesthesiologists following MDO was achieved in all patients with only a handful of subjects (3/63) continuing to have a difficult airway grade following MDO. Improved airway grade allows for easier intubation at future procedures/emergencies and is an approximation for improvement in baseline breathing. Tracheostomy was also avoided in all patients following MDO with most patients being discharged on room air.

Feeding outcomes were also encouraging as 80% of patients did not require a G-tube after MDO, 83% were taking some oral nutrition and 56% were taking full oral feeds at discharge. MDO allows for patients to be discharged with relatively low additional home care needs.

While nearly all patients remained below the 50th percentile for weight, growth curves remained relatively steady in the first year of life and the plateau seen on the growth curve prior to MDO resolved after surgery in both male and female patients. It should be noted that many of these patients had continued feeding challenges given their associated cleft palates that were not repaired until 10-12 months of age.

IMPLICATIONS

This study represents one of the largest cohorts of early MDO intervention. This investigation adds a wealth of patient data that support the efficacy of MDO in improving airway grade and maintaining growth. These results support the use of early MDO to allow for discharge with safe and relatively simple home care.

Future studies will include investigation into the feeding and objective sleep apnea outcomes using polysomnography following early MDO intervention. Future studies should focus on prospective investigations.

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DISCLOSURES

No relevant financial disclosures from any of the authors.