Curr Pediatr Res2023; 27 (05): 1-10

ISSN 0971-9032
www.currentpediatrics.com

Results of improving sleep of infants after MDO to treat severe Pierre robi sequence in Vietnam.

Dang Hoang Thom[1,3*], Vu Ngoc Lam[2], Tran Thiet Son[3]

[1]Department of Pediatrics, Vietnam National Children's Hospital, Hanoi, Vietnam

[2]Department of Nursing, 108 Military Central Hospitals, Hanoi, Vietnam

[3]Department of Plastic Reconstructive Aesthetic Surgery, Hanoi Medical University, Hanoi, Vietnam

Received: *26 April, 2023, Manuscript No. AAJCP-23-97378;* ***Editor assigned:*** *28 April, 2023, PreQC No. AAJCP-23-97378(PQ);* ***Reviewed:*** *09 May, 2023, QC No. AAJCP-23-97378;* ***Revised:*** *18 May, 2023, Manuscript No. AAJCP-23-97378(R);* ***Published:*** *29 May, 2023, DOI:10.35841/0971-9032.27.05.1185-1189.*

Abstract

Pierre Robin Sequence (PRS) refers to a group of birth defects typically include micrognathia and glossoptosis. These conditions can lead to airway obstruction at the base of the tongue and may be accompanied by cleft palate. Hypoplasia of the mandibular leads to glossoptosis, which results in obstruction of the upper airway, feeding difficulties and sleep disordered breathing. But there are no studies that have reported on the complete range of sleep-related outcomes in infants with Pierre Robin sequence that had undergone Mandibular distraction osteogenesis (MDO).

Objective: 73 infants aged 1-12 months with a diagnosis of Pierre Robin Sequence (PRS), who had undergone MDO (MDO) at the Vietnam national children's hospital between 2019 and 2021.

Methods: A longitudinal study.

Results: The 73 infants included, 32 (43.8%) were female, 11 (15.1%) were premature and 56 (76.7%) had isolated PRS. The average age at MDO was 35 days (IQR: 22 to 60 days). The infants were extubated on postoperative day 4 (IQR: 3 to 4 days). Tube feeding rate is 89.0%. The mean AHI of the subjects group decreased dramatically after MDO treatment; dropped from 25.5 pre-operatively to 1.7 post-operatively (p<0.001). Obstructive Apnea Hypopnea Index (AHI) decreased from 22.1 before MDO to 1.1 after MDO. The lowest SpO2 nadir increased from 74.3% to 84.2%, whereas obstructive apneas reduced from 76.2 to 0, hypopneas decreased from 48.1 to 22.1 (p=0.032), and time spent below 90% SpO2 decreased from 4.3% to 0.7% (p<0.001).

Conclusion: MDO improved several architectural parameters which strongly affect to sleep with PRS.

Keywords: Sleep, Pierre Robin, Jawbone, Infants.

Accepted on 12th May, 2023

Introduction

PRS is a group of birth defects including small jaw and tongue, with or without cleft palate; can lead to airway obstruction at the base of the tongue. In the United States, the incidence of PRS is quite low, ranging from 1 in 5000 to 1 in 7000 births [1-3].

Hypoplasia of the mandible leads to glossoptosis, which can lead to feeding difficulties, Sleep-Disordered Breathing (SDB), and obstruction of the upper airway [4-6]. SDB, especially in the form of Obstructive Sleep Apnea (OSA), is an almost universal finding in infants with PRS, with prevalence ranging from 85% to 100% [7]. The morbidity associated with OSA in infants is well described and this can result in failure to thrive, developmental and learning delays, cor pulmonale, and ultimately, death [8]. Costa, et al. found in a large retrospective review that mortality at PRS was 16.7% [9].

Recent publications advocate routine Polysomnography (PSG) screening in all patients with PRS who have difficulty eating and in patients with desaturations in the prone position [10,11]. The objective nature of PSG makes it an ideal tool to guide airway management and prevent Obstructive Sleep Apnoea (OSA) related morbidity [12,13].

The treatment of airway obstruction by Pierre Robin series has a wide range of options, from observation or prone position for mild cases to different types of surgery (including tongue-lip adhesions, osteogenesis) (mandible or tracheostomy) for severe cases [14,15]. MDO has become a popular treatment for airway obstruction in infants with Pierre Robin sequence that have micrognathia, as it directly addresses the underlying issue-the hypoplastic mandible [16]. Although there has been some limited evidence for resolution of obstructive sleep apnea following MDO. However, no study has reported comprehensive sleep-related outcomes in infants with PRS after MDO [17-19]. For that reason, our aim is to describe changes in sleep-related respiratory parameters in infants with Pierre Robin sequence that underwent MDO in Vietnam.

Methodology

We performed a longitudinal study of 143 infants aged 1-12 months diagnosed with PRS who underwent MDO at the Vietnam national children's hospital in the period of 2019 to 2021. The research was approved by Hanoi medical university review board.

We did not include children older than 12 months, lacking information in the medical records. The information on each subject was reviewed simultaneously by two independent Otolaryngologists for the results unifying. We conduct physical examination of all newborns; perform sleep endoscopy, direct laryngoscopy, rigid tube bronchoscopy and PSG.

The sleep endoscopy is the use of a flexible fiberoptic endoscope to examine the airway, tongue position, and jaw thrust when the infants were supine and supine. All patients were treated by the same team of surgeons. This team will be in charge of plastic and reconstructive surgery with mandibular distraction. Postoperative PSG was performed at the e[illegible]f the distraction process but before hardware removal.

General information on sex, age, and clinical sympt[illegible]ms rela[illegible] to upper airway obstruction were examined befor[illegible]d after MDO. FFL findings were documented in detail wit[illegible]ping of the tongue in various positions and findi[illegible]gs f lar[illegible] palsy. Classification pediatric otolaryngolo[illegible]ts will classify laryngomalacia according to the mild, mo[illegible]te and severe grades associated with epiglottitis coll[illegible] Du[illegible]physical examination at the time of Flexible Fi[illegible]tic Laryngoscopy (FFL) and direct laryngoscopy a[illegible]noscopy, the additional signs of reflux as [illegible]as [illegible]way compromise were also noted. Regarding th[illegible]ethod o[illegible]ata processing, we employ a paired sample t-[illegible]that was used to compare the pre- and post-MDO [illegible]eans of [illegible]ement before and after the MDO.

Results

[illegible]ohort [illegible]3 in[illegible]nts with PRS, 109 (76.2%) treated by airw[illegible]related s[illegible]y (either MDO or tracheostomy).

Sev[illegible]nfants (66.9%) were eventually included in the [illegible]y because they met all inclusion criteria There were 36 [illegible] who were not eligible to participate in the study due to: crit[illegible] for exclusion included being intubated at birth during polysomnography, undergoing polysomnography more than 3 months after MDO, and having undergone oxygen titration or a split-night study with oxygen. Of the 73 infants included, 32 (43.8%) were male, 11 (15.1%) were premature and 56 (76.7%) had isolated Pierre Robin sequence (Table 1).

Demographic and clinical characteristics	All infants (%)
No.	73
Male	32 (43.8)
Prematurity	11 (15.1)
Isolated PRS	56 (76.7)
Age at MDO, day	
Median	35
IQR	22-60
Post-operative intubation, days	
Median	4
IQR	[illegible]-4
Total hospital lengt[illegible]	
Median	29
IQR	23-39
Tube feeding ([illegible]G or GT) at pre-MD[illegible]	65 (89.0)
Note: NG[illegible]Naso[illegible]Tube; GT: Gastrostomy Tube; IQR: Interquartile Range	

Table 1. [illegible]ographic and clinical characteristics of infants.

The [illegible]rage [illegible]ge of MDO patients was 35 days (interquartile [illegible]nge, [illegible] to 60 days). The median age at MDO was not [illegible]ific[illegible]ntly different between infants with isolated Pierre Robin Sequence and those with syndromic Pierre Robin Sequence (36 days (interquartile range, 22 to 60) *vs.* 29 days (interquartile range, 21 to 85 days)).

No postoperative complications occurred and the children were extubated on postoperative day 4 (interquartile range, 3 to 4 days). The length of hospital stay was 29 days (interquartile range, 23 to 39 days). Tube feeding rate is 89.0%.

Some aspects of sleep architecture were discovered to improve following surgery. The individuals' mean AHI fell significantly after MDO treatment. The mean AHI fell from 25.5 pre MDO to 1.7 post MDO, a mean difference of 23.8 ($p<0.001$); obstructive AHI decreased from 22.1 before MDO to 1.1 after MDO, both of which were substantial improvements.

The lowest SpO2 nadir increased from 74.3% to 84.2%, whereas obstructive apneas reduced from 76.2 to 5.8, hypopneas decreased from 48.1 to 22.1 ($p=0.032$), and time spent below 90% SpO2 decreased from 4.3% to 0.7% ($p<0.001$). No infants have required tracheostomy as part of their airway treatment after surgery (Figures 1 and 2).

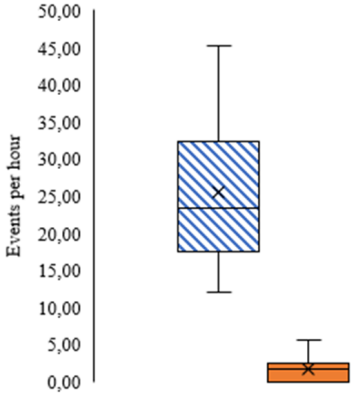

Figure 1. Comparison of Apnea Hypopnea Index (AHI) before and after MDO. **Note:** AHI pre MDO: ( ); AHI post MDO: ( ).

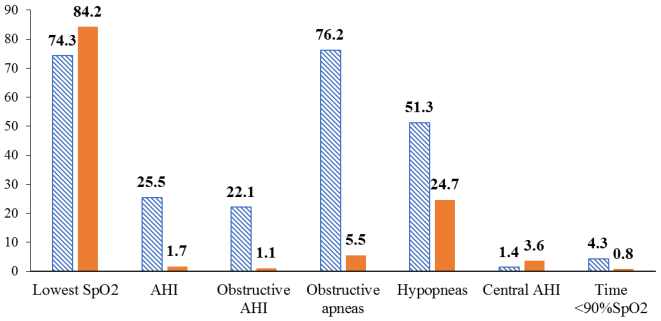

Figure 2. The difference of various aspects of sleep architecture between pre MDO and post MDO. **Note:** Pre MDO (▨); Post MDO (■).

Discussion

MDO was first applied for infant's treatment with hemifacial microsomia and treacher-collins syndrome in the 1990's [20-22]. MDO is a less invasive option to procedures that have a higher morbidity and complication rate. Then, plenty reports have been published which demonstrating the procedure's efficiency and safety [23-25]. According to the literature, the MDO reduces the severity of OSA by preventing the need for a tracheostomy, assisting with decannulation, and reducing the need for tracheostomy [26-28].

The MDO expands the infant airway's anterior-pos or dimensions by lengthening the mandible and shifting th geniohyoid muscle attachment anteriorly. Lam, et al. demonstrated in a retrospective cohort study that M can prevent tracheostomy in PRS patients and te decannulation in previously tracheostomised pati s [29]. overall success rate in this study was 75.6%. eanwhile, th lower complication rate, only 26.8%, was crit y highe in the group of tracheostomy than in the M oup. Breik, et al. performed a 2016 meta-st dy 66 articles that showed the success rate (defined as preve ti heostomy) of 95.5% [26].

Among the patients in this the mean AHI significantly decreased from 25.5 eo erati to 1.7 postoperatively, with a mean difference of 8 ($p<0.$ The mean Obstructive Apnea-Hypopnea Index HI) d creased from 22.1 to 1.1 after surgery. Th an lo O_2 saturation nadir increased from 74.3% to 4.2 . The SpO2 dropped from 4.3% to 0.8%. The sults a with e data from earlier sources. In the udy Papo n. the author evaluated the treatment esults o 24 PSG patients before and after MDO surgery. The ly ved that the average AHI of this group of s s decreased from 47 to 10.9 after 1 month and further decr d to only 2.5 after 1 year. The mean lowest value of O_2 saturati increased from 76.5% to 98.3% after 1 month and continued to increase to 98.5% after 1 year from the time of surgery.

This study also compared these results with a control group undergoing TLA and presented that MDO was dominant to TLA in patients without the syndrome [30]. Briek, et al. in their study found 11 articles comparing obstructive AHI before and after surgery. As a result, AHI was significantly reduced from 31.2 (before surgery) to 4.34 (after surgery), with a mean difference of 26.9 [26]. Another large retrospective study of 121 patients reported significantly reduced mean AHI across all weight groups. However, children weighing<4 kg had better reductions than children weighing>4 kg (41.5 *vs.* 26.1).

The study concluded that MDO is an effective t tment method for low birth weight infants with hig ety a ow complications. To further elucidate the relati ip bet sleep indices and clinical symptoms of neon disease, Daniel, et al. performed a recent revie e lit e on neonatal PSG [31]. The autho studied 0 pap n the values of various parameters of PS infant he re ts show that the upper limit of normality for C HI a AI is reported to be less than 1.0. In c a gradual decrease with age in the central api index er limit was 45 for 1 month old infants, 30 fo 2 month olds, 22 for 3 month olds, and 10-20 for elder age grou s. The change in central apnea in the growing infant is thought be due to the variation in respiratory ph logy from the fetal to the infant's period.

Conclu n

In our re h sample of infants with PRS, MDO d me ted t achievement well increasing parameters of ep, in ticular AHI. By the study of 73 children with PRS s ome, mandibular stretching is highly effective in imp g jawbone size and increasing airway size for diatric patients. Thereby, it results in improving respiratory gestion and reducing the difficulty of nursing. The limitation of the study is that the number of infants who completely performed PSG after 3 months of MDO was still low compared to the total number of infants treated. Inter-observer and internal reliability is ensured through data collection. All steps were performed exactly as described in the methods.

Conflicts of Interest

All authors in this study have no conflicts of interest, financial or otherwise.

References

1. Scott AR, Mader NS. Regional variations in the presentation and surgical management of Pierre Robin sequence. Laryngoscope 2014; 124(12): 2818–25.
2. Hsieh ST, Woo AS. Pierre robin sequence. Clin Plast Surg 2019; 46(2): 249–59.
3. Giudice A, Barone S, Belhous K, et al. Pierre robin sequence: A comprehensive narrative review of the literature over time. J Stomatol Oral Maxillofac Surg 2018; 119(5): 419–28.
4. Daniel M, Bailey S, Walker K, et al. Airway, feeding and growth in infants with Robin sequence and sleep apnoea. Int J Pediatr Otorhinolaryngol 2013; 77(4): 499–503.
5. Anderson ICW, Sedaghat AR, McGinley BM, et al. Prevalence and severity of obstructive sleep apnea and

snoring in infants with Pierre robin sequence. Cleft Palate Craniofac J 2011; 48(5): 614–8.
6. Gómez OJ, Barón OI, Peñarredonda ML. Pierre robin sequence: An evidence-based treatment proposal. J Craniofac Surg 2018; 29(2): 332–8.
7. Cladis F, Kumar A, Grunwaldt L, et al. Pierre robin sequence: A perioperative review. Anesth Analg 2014; 119(2): 400–12.
8. Singer LP, Saenger P. Complications of pediatric obstructive sleep apnea. Otolaryngol Clin North Am 1990; 23(4): 665–76.
9. Costa MA, Tu MM, Murage KP, et al. Robin sequence: Mortality, causes of death, and clinical outcomes. Plast Reconstr Surg 2014; 134(4): 738–45.
10. Reddy VS. Evaluation of upper airway obstruction in infants with Pierre robin sequence and the role of polysomnography–Review of current evidence. Paediatr Respir Rev 2016; 17: 80–7.
11. Zaballa K, Singh J, Waters K. The management of upper airway obstruction in Pierre robin sequence. Paediatr Respir Rev 2022; S1526-0542(22): 00047-1.
12. Dennison WM. The Pierre robin syndrome. Pediatrics 1965; 36(3): 336–41.
13. Insalaco LF, Scott AR. Peripartum management of neonatal Pierre robin sequence. Clin Perinatol 2018; 45(4): 717–3
14. Perrino MA. Neonatal mandibular distraction. Atlas Oral Maxillofac Surg Clin North Am 2022; 30(1) 62.
15. Rossini G, Vinci B, Rizzo R, et al. Mandibular distraction osteogenesis: A systematic review of stability and the effects on hard and soft tissues. Int J Oral Maxillofac Surg 2016; 45(11): 1438–44.
16. Hong P, Bezuhly M. Mandibular distraction osteogenesis in the micrognathic neonate: A review for neonatologists and pediatricians. Pediatr Neonatol 2013; 54(3): 153–60.
17. Côté A, Fanous A, Almajed A, et al. Pierre robin sequence: Review of diagnostic and treatment challenges. Int J Pediatr Otorhinolaryngol 2015; 79(4): 451–64.
18. JC, Br JP. Surgical considerations in Pierre robin sequence. Clin Surg 2014; 41(2): 211–7.
19. Me GG, Kaur N, Hura KS. Pierre robin sequence: Diagnostic difficulties faced while differentiating isolated and syndromic forms. Acta Medica (Hradec Kralove). 2 3(2): 86–90.
20. McCarthy JG. The role of distraction osteogenesis in the reconstruction of the mandible in unilateral craniofacial microsomia. Clin Plast Surg 1994; 21(4): 625–31.
21. Moore MH, Guzman-Stein G, Proudman TW, et al. Mandibular lengthening by distraction for airway obstruction in treacher-collins syndrome. J Craniofac Surg 1994; 5(1): 22–5.
22. Aljerian A, Gilardino MS. Treacher collins syndrome. Clin Plast Surg 2019; 46(2): 197–205. [Crossref][Google Scholar][Indexed]
23. Earley M, Butts SC. Update on mandibular distraction osteogenesis. Curr Opin Otolaryngol Head Neck Surg 2014; 22(4): 276–83.
24. Hatefi S, Hatefi K, Le Roux F, et al. Review of automatic continuous distraction osteogenesis devices for mandibular reconstruction applications. Biomed Online 2020; 19(1): 17.
25. Starch-Jensen T, Viellerup AB, Jæhr TL. Mandibular midline distraction osteogenesis with a bone-borne, tooth-borne or hybrid distraction appliance: A systematic review. J Oral Maxillofac Res 2018; 9(3): e1.
26. Breik O, Grey D, Umapathysivam K, et al. Mandibular distraction osteogenesis for the management of upper airway obstruction in children with micrognathia: A systematic review. Int J Oral Maxillofac Surg 2016; 45(6): 22.
27. Nin Landoval TC, Rodrigues EDR, Vasconcelos BC. Latency phase in mandibular distraction osteogenesis: A systematic review in animal models. Br J Oral Maxillofac Surg 2021; 59(9): 993–1004.
28. Shakir S, Bartlett SP. Modern mandibular distraction applications in hemifacial microsomia. Clin Plast Surg 2021; 48(3): 375–89.
29. Lam DJ, Tabangin ME, Shikary TA, et al. Outcomes of mandibular distraction osteogenesis in the treatment of severe micrognathia. JAMA Otolaryngol Neck Surg 2014; 140(4): 338–45.
30. Papoff P, Guelfi G, Cicchetti R, et al. Outcomes after tongue–lip adhesion or mandibular distraction osteogenesis in infants with Pierre robin sequence and severe airway obstruction. Int J Oral Maxillofac Surg 2013; 42(11): 1418–23.
31. Daniel KN, Chan CH. A review of normal values of infant sleep polysomnography. Pediatr Neonatol 2013; 54(2): 82–7.

***Correspondence to:**

Dang Hoang Thom

Department of Plastic Reconstructive Aesthetic Surgery,

Hanoi Medical University,

Hanoi, Vietnam

E-mail: drthomdh@gmail.com