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Surgical management and outcomes of Pierre Robin Sequence: a comparison of mandibular distraction osteogenesis and tongue-lip adhesion

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Abstract

There is a paucity of primary literature directly comparing tongue-lip adhesion (TLA) versus mandibular distraction osteogenesis (MDO) in surgical treatment of patients with Pierre Robin Sequence (PRS). This study comprehensively reviews the literature to evaluate and compare the effectiveness of MDO and TLA in improving airway and feeding outcomes. A search was performed using the MEDLINE (PubMed interface) and Embase databases for publications between 1960 through June 2017. English-language, original studies involving MDO or TLA in treatment of PRS subjects were included. Extracted data included prevention of tracheostomy (primary airway outcome) and ability to feed exclusively by mouth (primary feeding outcome). Sixty-seven studies total were included in the review. Ninety-five percent (657/693) of subjects treated with MDO avoided tracheostomy. Eighty-nine percent (289/323) of subjects treated with TLA avoided tracheostomy. Eighty-seven percent (323/370) of subjects treated with MDO achieved full oral feeds at latest follow-up. Seventy percent (110/157) of subjects treated with TLA achieved full oral feeds at latest follow up. The rate of second intervention for recurrent obstruction ranged from 4 to 6% in MDO studies, compared to range of 22 to 45% in TLA studies. Variability of patient selection, surgical techniques, outcomes measurement methods, and follow up length across studies precluded meta-analysis of the data. Both MDO and TLA are effective alternatives to tracheostomy for patients who fail conservative management, improve feeding and promote weight gain. MDO may be superior to TLA in long-term resolution of airway obstruction and avoidance of gastrostomy, but is associated with notable complications.

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IRB:
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Introduction

Pierre Robin sequence (PRS), first described in the French literature in 1934[1], refers to the triad of micrognathia, glossoptosis and airway obstruction (Figure 1). In 73–90% of cases[2–5], infants also present with a U-shaped or V-shaped cleft of the secondary palate. The incidence reported in the literature ranges from 1/3120[6] live births to 1/8060 live births[4]. PRS can be an isolated finding, or can be associated with a genetic syndrome in approximately 50% of cases[2,7–10].

The two primary clinical sequalae of PRS are tongue-based airway obstruction and feeding difficulties. The small jaw contributes to the tendency for the tongue to fall back into the pharynx, occluding the upper airway[11]. The airway obstruction, as well as the associated cleft palate, contributes to poor weight gain and feeding difficulties. An estimated 40–70% of PRS cases need nasogastric tube feeding for up to several months, and may even require gastrostomy tube feeding[12–14].

Treatment of airway obstruction begins with non-invasive techniques. Placing the infant in a prone position allows for the tongue to fall forward and reduce airway obstruction. In 40–70% of cases, positioning alone is successful in relieving obstruction[12,15,16]. If positioning fails to relieve airway obstruction, placement of a nasopharyngeal airway (NPA) can serve as a temporizing measure while awaiting mandibular growth[17,18]. Infants requiring additional respiratory support may require continuous positive airway pressure (CPAP)[19].

Although conservative management is sufficient to relieve airway obstruction for many neonates with PRS[20], those who fail will ultimately require surgical intervention. Historically, tracheostomy was the only option for long-term airway stabilization[21] and the preferred intervention among otolaryngologists[22]. However, it is associated with significant morbidity and mortality[23]. Carr et al[110] reported a 43% rate of serious complications and 0.7% mortality rate, mostly due to accidental decannulation or obstructions. Tracheostomy is burdensome for caretakers[111], requiring regular maintenance and suctioning to prevent mucous obstruction[112]. 60% of parents of children with non-syndromic and 80% of parents of children with syndromic PRS surveyed described the overall experience with tracheostomy as difficult and cited prolonged tracheostomy as a major concern[111]. Furthermore, children with PRS who underwent tracheostomy were not decannulated until an average age of 3.1 years[21]. The prolonged dependence on tracheostomy may contribute to significant delays in speech production and language development[113].

Previously, the only surgical alternative to tracheostomy was glossopexy or tongue-lip adhesion (TLA). Popularized by Douglas in 1946[24], with subsequent revisions in technique[25–27], the procedure anchors the tongue to the lower lip and mandible, securing the tongue in an anterior lingual position, preventing occlusion of the upper airway (Figure 2). It is typically performed in the first months of life and reversed at around 12 months of age, often at the time of cleft palate repair.

More recently, mandibular distraction has gained popularity as an alternative method of treating this form of airway obstruction. Initially applied to patients who had already undergone tracheostomy and/or TLA[28,29], over the past decade it has been adopted by many centers as the primary surgical intervention for tongue-based airway obstruction in PRS patients. Lengthening the mandible brings the tongue forward through its attachments to the lingual surface of the mandible, addressing both glossoptosis and micrognathia[30], relieving occlusion of the airway and creating more space for the tongue (Figure 3) (See Video, Supplemental Digital Content 1, which demonstrates our technique for mandibular distraction osteogenesis for patients with PRS, available in the "Related Videos" section of the Full-Text article on PRSJournal.com or, for Ovid users, available at INSERT HYPER LINK) (Video Graphic 1).

In addition to avoiding the complications associated with tracheostomy, both MDO[114–116] and TLA[6] have a lower cost burden. However, it is important to emphasize that tracheostomy may be the optimal and life-saving intervention for patients with multiple sites of airway obstruction (i.e. subglottic stenosis, tracheomalacia), significant secondary respiratory abnormalities and central apnea[85,109].

The decision to undergo either MDO or TLA is influenced greatly by the surgeon's specialty and experience[7,22,31–33]. A number of studies have proposed algorithms and patient selection guidelines[34–39], but these rely largely on expert opinion or results from small studies. Furthermore, there is a paucity of primary literature directly comparing outcomes of TLA versus MDO.

Given the limited number of original research studies, this review aims to comprehensively compile data on specific defined endpoints from the existing literature. Evaluation of the data on airway and feeding outcomes in MDO and TLA will assist surgeons caring for PRS patients with their choice of intervention and in counseling of families.

Methodology

A search was performed using the MEDLINE (PubMed interface) and Embase databases for publications between 1960 through June 2017. Selected articles were analyzed with regard to level of evidence[40], patient demographics, length of follow up, diagnosis, distractor type, airway and feeding outcomes. With the exception of two studies[41,42], all studies included a mixed cohort of syndromic and isolated PRS subjects. We present our search criteria in Table 1, and literature selection and outcomes measurement methodology in Figure 4.

Results

Airway outcomes and feeding outcomes extracted from individual studies are presented in Table 2 and Table 3, respectively, along with pertinent patient demographics and reported complications. Table 4 summarizes the aggregated data for the primary outcomes, specifically avoidance of tracheostomy and achievement of full oral feeds at latest follow up. Given that most studies were single intervention case series that did not directly compare MDO and TLA, the four comparison studies, the highest level of evidence available in the literature, are presented in detail in Table 5.

Airway Outcomes

Avoidance of tracheostomy:

Ninety-five percent (657/693) of subjects treated with MDO avoided tracheostomy. Eighty-nine percent (289/323) of subjects treated with TLA avoided tracheostomy. In a large MDO cohort study (34% syndromic), tracheostomy was avoided in 93% (76/81) of subjects after 22 months of follow up[106]. Tracheostomy was avoided in 89% of a large TLA cohort study (45% syndromic)[36]. One hundred percent of those treated with MDO, versus 73% of those treated with TLA, avoided tracheostomy in a retrospective historical comparison study of an isolated PRS cohort with at least 1 year of follow up (no p value provided)[42]. All subjects in both the MDO and TLA cohorts avoided tracheostomy in a prospective, nonrandomized study with at least one year of follow up[43].

Decannulation:

Eighty percent (165/206) of subjects achieved decannulation following MDO. Notably, decannulation was achieved in only 17% (2/12) of a cohort where all subjects had complex congenital syndromic comorbidities[44]. However, decannulation was achieved in 100% of a cohort (11/11) that was 72% syndromic and had severe OSA[46].

Relief of OSA:

Both a prospective nonrandomized study[43] and a retrospective historical comparison study[42] found that MDO subjects had significantly lower postoperative apnea-hypoapnea index (AHI) compared to TLA subjects, despite having significantly higher AHI preoperatively.

Improvement in oxygen saturation:

After MDO, oxygen saturation levels improved from a preoperative range of 72–76% to a postoperative range of 91–95%[28,45–47]. After TLA, oxygen saturation levels increased from a preoperative range of 63–73% to a postoperative range of 81–88%[48–50]. A retrospective historical comparison study found that patients undergoing MDO had significantly higher oxygen saturation levels than TLA patients both at 1 month (98.3 percent versus 87.5 percent; $p < 0.05$) and 1 year postoperatively (98.5 percent versus 89.2 percent; $p < 0.05$) despite having higher preoperative AHI[42].

Repeat procedure:

Denny et al reported that 45% of patients (5/11) treated with TLA eventually required a secondary surgery for recurrent airway obstruction (four distractions and one repeat TLA)[51]. Three other studies reported 22% (2/9)[56], 27% (3/11)[2], and 29% (4/14)[52] of subjects required repeat procedure due to early disruption from wound breakdown. In contrast, the rates of repeat distraction were 4% (1/23)[53], 5% (1/19)[54], and 6% (3/50)[55].

Feeding Outcomes

Achievement of full oral feeds:

Eighty-seven percent (323/370) of subjects treated with MDO achieved full oral feeds at latest follow up. Eleven percent (42/370) still required gastrostomy for supplemental feeding at latest follow up. By comparison, 70% (110/157) of subjects treated with TLA achieved full oral feeds at latest follow up, with 20% (32/157) requiring gastrostomy for supplemental feeding. A retrospective historical comparison found that subjects undergoing TLA were 6.5 times more likely to require a gastrostomy tube for nutritional support, compared to subjects undergoing MDO[57].

Of note, in a single-center retrospective study where all infants achieved full oral feeds, infants resumed oral feeding sooner after MDO than after TLA, despite the MDO cohort containing significantly more syndromic patients[56]. However, no significant difference in time needed to discontinue tube feeds was found in a prospective nonrandomized study[43].

Growth/weight gain:

While most MDO studies[53,58–60] reported improved weight gain following distraction, one study[61] found an early decline in growth rate in 7 out of 10 subjects that persisted for 12 months. Cozzi et al found that infants had an increase in both body weight percentile and weight velocity percentile following TLA[62]. In contrast, Bijnen et al reported that 11 of the 21 subjects remained within the same growth percentile[63]. No significant difference in growth between MDO and TLA patients was found both in a prospective, nonrandomized study[43].

Improvement in gastroesophageal reflux (GER):

No studies reviewed specifically reported on GER before and after TLA. Hong et al reported that all 6 subjects who underwent MDO had resolution of GER that was seen on preoperative upper GI series, and were able to discontinue anti-reflux medical therapy[65]. Similarly, Genecov et al found that only 3% of patients had GER postoperatively, a decrease from 67% with GER preoperatively[66].

Swallow function:

Two MDO studies[45,65] specifically evaluated swallowing function following surgical intervention. Both studies reported resolution of laryngeal penetration (passage of materials into the larynx) and aspiration (passage of materials through the vocal folds) following distraction.

Complications

While not the main outcome of comparison in this review, the relative risks associated with either intervention should be noted. Perioperative complications reported in individual studies are presented in Tables 2 and 3.

Complications of MDO include external scarring[85], hypertrophic scarring[101], infection[45,47,65,80,89,], hardware exposure[93], device dislodgment or pin loss[28,90,98,99,102], facial nerve problems[101,47,66] and tooth-bud damage[101]. Tibesar et al retrospectively reviewed and reported open bite deformity (28%), dental complications (16%) and facial nerve injuries (9%) as the leading complications in the long-term period[54]. While TLA causes minimal external scarring compared to MDO, its complications include wound dehiscence[34,48,51,52], abscesses and mucoceles[63,108], and injuries to Wharton's ducts[51,63].

Discussion

TLA and MDO, both adopted as alternatives to tracheostomy, have distinct inherent advantages and disadvantages. These features are presented in Table 6, along with the most commonly cited complications. TLA is a relatively simple procedure and provides immediate relief of obstruction by moving the tongue. However, the procedure is only a temporizing measure that relies on "catch-up growth" of the mandible to ultimately relieve airway obstruction. While some have reported subsequent normal growth of the mandible[35,67], one study found various structures did not reach normal values compared to controls[68]. Certain cases of syndromic PRS, including Treacher Collins and hemifacial microsomia, have persistent micro/retrognathia compared to isolated PRS, Stickler or velocardiofacial syndrome[69].

MDO corrects micrognathia, in contrast to TLA, by lengthening the mandible and increasing mandibular volume[29,70]. But the procedure is technically more difficult and requires surgeons with specialized training and equipment, necessitates a secondary operation for removal of hardware, and is associated with complications such as hardware infection, dental damage and nerve injury. Furthermore, improvement of the airway obstruction is gradual, as the mandible is slowly lengthened through distraction. Thus, the airway may require monitoring in an intensive care setting during the distraction period.

Prior reviews

Several reviews have been published on the management of PRS, MDO and TLA. A systematic review by Viezel-Mathieu et al[71] found TLA was successful in relieving airway obstruction in 81.3% patients. Our review of the literature yielded a greater success rate of 89% in TLA subjects. However, we specifically defined the primary endpoint as avoidance of tracheostomy. A recent review by Tahiri et al.[72] found that MDO successfully treated airway obstruction in 89.3% of cases. Ow and Cheung conducted a meta-analysis of MDO studies published between 1966–2006[73]. Their analysis yielded a 91% rate of prevention of tracheostomy and a 78% rate of decannulation. Our findings (95% prevention of tracheostomy and 90% decannulation) may differ due to inclusion of more recently published studies, and inclusion of only subjects with diagnosis of PRS. Finally, Breik et al.[74] found that MDO led to 82% of children feeding exclusively orally after surgery, and resolution of GERD in 66/70 patients. Our review found that 87% of subjects reported in the literature achieved exclusive oral feeding. We included subjects who initially required supplemental feeding but later discontinued at latest follow up, which may contribute to our higher rate.

Few systematic reviews have included both TLA and MDO interventions. Bookman et al. [75] conducted a systematic review on neonates with tongue-based airway obstruction, and found a lack of consistency in diagnosis, treatment protocols and reporting of outcomes. The review did not include more recent prospective studies or studies comparing both MDO and TLA patients. Almajed et al[76] systematically compared reported PSG outcomes following MDO and TLA. MDO was associated with the lowest percentage of significant airway obstruction postoperatively (3.6%) compared to 50% for infants who underwent TLA. No prior reviews have directly compared feeding outcomes following TLA versus following MDO.

Airway Outcomes

TLA is effective for relief of severe upper airway obstruction, with 89% of subjects reported in the literature being able to avoid a tracheostomy. Similarly, tracheostomy was prevented in 95% of MDO subjects. Furthermore, MDO can improve airway obstruction even in patients who previously required tracheostomy, with successful decannulation in 80% of subjects. Aggregate data from the literature reviewed and comparison studies suggest that MDO promotes greater resolution of OSA and higher oxygen saturation, compared to TLA. MDO may provide more stable and long-term relief, as a higher percentage of infants treated with TLA required a repeat procedure for recurrent airway obstruction.

Feeding Outcomes

Both TLA and MDO can relieve feeding difficulties, with 70% and 88% of subjects, respectively, resuming full oral feeds. Improved feeding function promotes continued weight gain and growth following MDO and TLA, although infants may continue to be at lower weight percentiles. No significant difference in long-term weight gain or growth was found between MDO and TLA[64]. However, the higher percentage of MDO subjects achieving full oral feeds seen in the aggregate data, as well as findings from two retrospective comparison studies[56,57], suggest that MDO has an advantage over TLA with earlier return to oral feeding and discontinuation of supplemental feeding, and lower rates of gastrostomy placement. The avoidance of an additional surgical intervention and decreasing time needed to care for a feeding tube are important potential benefits for patient quality of life and caretaker burden. Moreover, although limited studies are available, MDO may also promote normal feeding by relieving GER and improving swallowing function.

Recommendations for clinical practice

Regardless of the intervention, careful patient selection is necessary to anticipate potential complications and achieve optimal outcomes. In both MDO[53,60,61,77] and TLA[36,48,78] syndromic patients tended to have poorer airway and feeding outcomes. Subjects who still required gastrostomy after either MDO or TLA often had an associated syndrome [43,53,56,60]. Poorer odds of success of tracheostomy avoidance or decannulation with MDO were associated with having a diagnosis of craniofacial microsomia or Goldenhar syndrome (OR, 0.07 [95% CI, 0.009–0.52])[39]. Syndromic diagnosis is similarly associated with an increased risk of failure of TLA, in addition to gastroesophageal reflux disease, preoperative intubation, late surgical intervention, and low birth weight (GILLS acronym)[36]. Gastroesophageal reflux and age >30 days were also associated with failure of MDO to avoid tracheostomy[79]. Definitive conclusions regarding the choice of intervention for PRS with associated syndromes or comorbidities cannot be drawn from this review.

In several instances, patients who failed initial intervention, ultimately requiring tracheostomy, were found to have concomitant airway anomalies, such as laryngomalacia, tracheal webs and vascular rings[36,48,52,79–83]. Patients with central apnea or neurologic disabilities also were likely to fail either MDO or TLA[36,56,66,79,84,85]. Most studies excluded patients with these comorbidities from their analysis, although one study on subjects with layrngomalacia treated with MDO reported 100% success in avoidance of tracheostomy or decannulation[86]. Thorough preoperative airway evaluation (i.e. nasopharyngoscopy or bronchoscopy to identify other causes of obstruction other than glossoptosis[42,43]) and polysomnogram[62], as well as assessment of other comorbidities suggesting a syndrome, allows for the most appropriate selection of intervention. The results of this review suggest that patients with laryngomalacia may still be candidates for MDO, but TLA should be avoided[86,87]. Furthermore, patients with significant lower airway anomalies, who are likely to fail either MDO or TLA, may preferably undergo tracheostomy primarily[34,85].

Future Directions

Despite the possible advantages MDO may have over TLA in long-term relief of airway obstruction and feeding difficulties, it has the notable disadvantage of requiring a second operation for removal and complications such as nerve and tooth bud damage. Additional studies are required to identify risk factors for MDO perioperative complications. Of particular interest would be whether syndromic PRS subjects are more prone to perioperative complications, given that several studies have shown they have poorer feeding and airway outcomes[39,53,60,61,77]. Expanded knowledge of risk factors and continued advances in operative technique can help to reduce complications, increasing the benefit to risk ratio of MDO over TLA.

The comprehensive literature search performed for this review yielded primarily retrospective, lower-level evidence studies (case series and cohort studies). Only five studies compared outcomes of MDO and TLA subjects, of which one study was prospective[43]. Parents were offered both interventions, but the authors suggest that surgeons may have unintentionally counseled parents of neonates with more severe phenotypes towards MDO, given that the MDO cohort had more severe baseline characteristics than the TLA cohort. In all four retrospective comparisons[42,56,57], all TLA subjects were treated prior to the adoption of MDO at the institution. The results may be confounded by other factors that may have changed between the beginning and end of the study period.

Before reaching a consensus on optimal treatment modality for PRS patients, more carefully designed prospective studies comparing the two interventions need to be conducted. These studies should institute a standardized protocol for defining and diagnosing PRS, preoperative evaluation to assess for comorbidities, and objective measurements of short and long-term airway function. Subjects should be carefully followed for nutritional requirements and growth.

Limitations

This review assesses the best evidence available at the time of publication to compare airway and feeding outcomes of MDO and TLA subjects. However, the ability to rigorously compare outcomes was limited by variations in the selection of cohorts, and the severity of phenotype. Most studies included mix of syndromic and nonsyndromic isolated PRS. The majority excluded patients with central apnea, neurological disabilities and/or concomitant airway anomalies, but a few included these potentially higher risk patients. Surgical techniques also varied between institutions, which could affect rate of surgical complications and success in improving outcomes. Studies varied in length of follow up. We are not able to account for the possibility of higher success rates simply because some cohorts were followed for shorter periods of time, before patients could develop recurrence in airway obstruction. Finally, the lack of standardization in reporting outcomes after surgical intervention creates difficulties in direct comparisons of MDO and TLA outcomes. The strength of this review is that it included only studies where defined endpoints could be extracted. Furthermore, we have highlighted variations in patient selection, technique and methods of reporting outcomes, as well as level of evidence grading, in summary tables of individual studies.

Conclusion

A comprehensive review of the literature regarding surgical management of PRS and airway and feeding outcomes has led to several determinations: 1) Both MDO and TLA are effective alternatives to tracheostomy for patients who fail conservative management 2) Both interventions improve feeding and promote weight gain 3) MDO may be superior to TLA in long-term resolution of airway obstruction and avoidance of gastrostomy. 4) MDO has a higher rate of reported long-term complications, including scarring, dental damage and facial nerve injuries. 5) Definitive conclusions on which intervention yields better outcomes cannot be made due to variability of patient selection, surgical techniques, and follow up length.

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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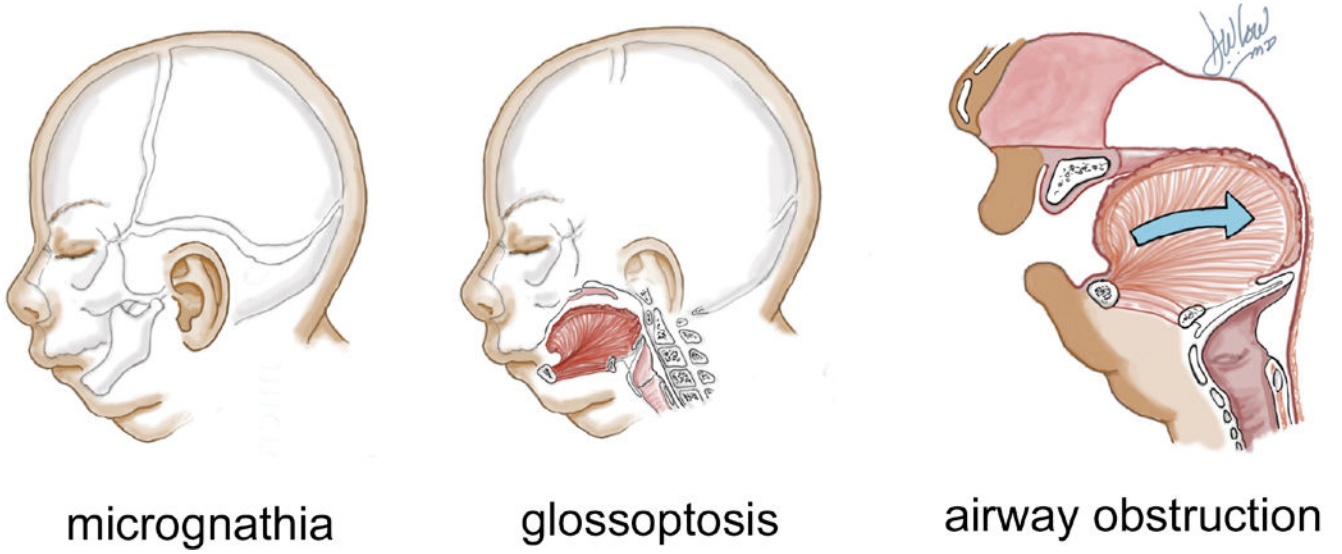

Figure 1.
Pierre Robin Sequence triad: micrognathia, glossoptosis, upper airway obstruction

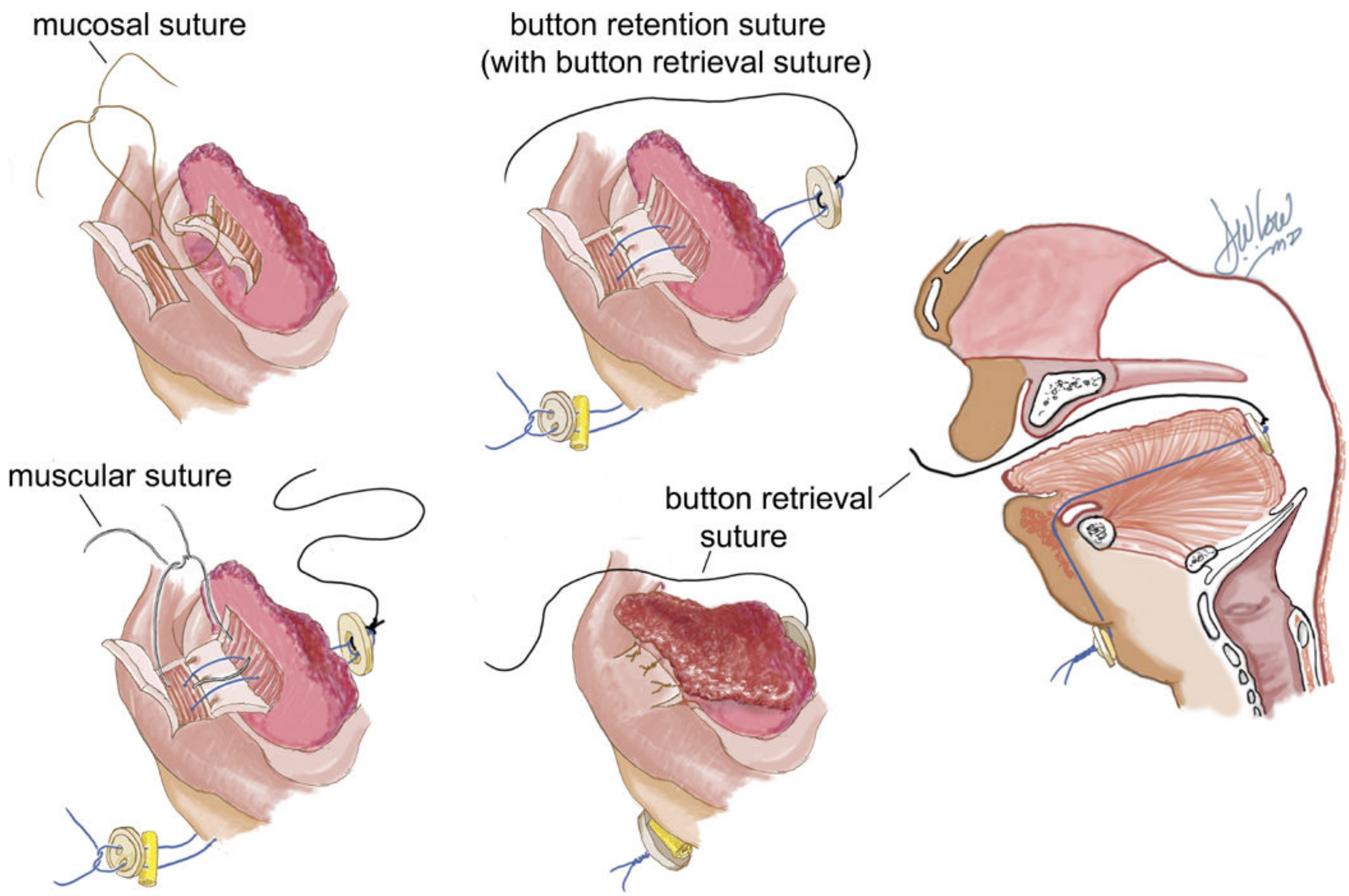

Figure 2.
Tongue-lip adhesion technique showing mucosal, button retention, muscular and button retrieval sutures

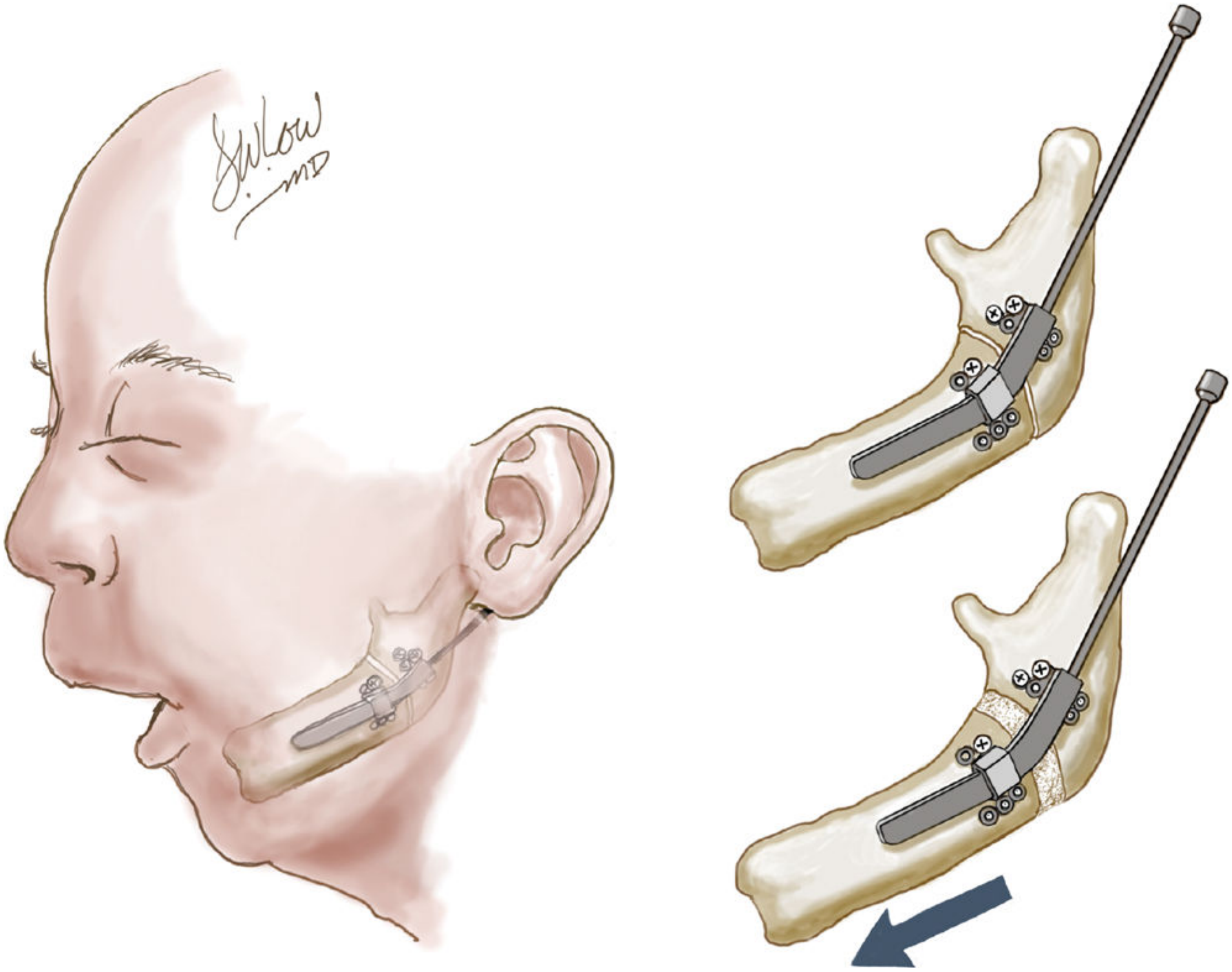

Figure 3.
Mandibular distraction osteogenesis in the micrognathic patient.

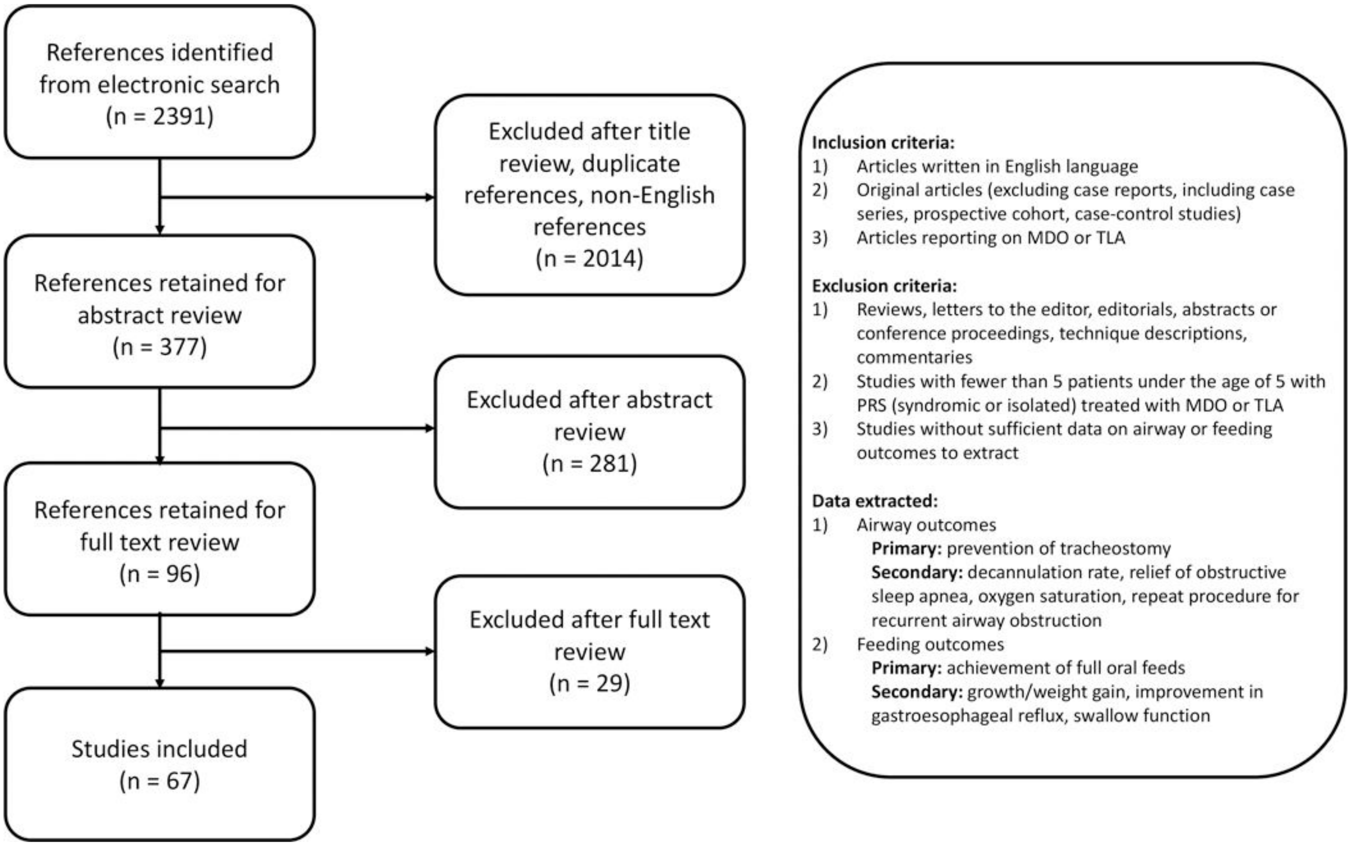

Figure 4.
Flowchart of literature selection criteria and strategy

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TABLE 1.

Database and search terms used to identify relevant articles

Database	Date Searched	Search Terms
Medline	1960 to June 2017	MeSH terms: “Pierre Robin syndrome”, “airway obstruction” combined with “retrognathia” or “micrognathia”, “osteogenesis, distraction” combined with “airway obstruction”; Free text keywords: “glossopexy”, “tongue lip adhesion”
Embase	1960 to June 2017	Disease search term: “pierre robin syndrome” queried with subheadings “surgery”, “disease management” and “therapy”

Table 2.

Comparison of Airway Outcomes

MDO article	Type of Distractor	Patient selection/surgical indication	Tracheostomy prevented /Decannulation	OSA	Perioperative Complications	Follow up length	Level of Evidence/ MINORS	Notes
Al-Samkari HT et al. 60	external	42% syndromic, failure of conservative management	Prevented: 91.67% (11/12)	N/A	Not reported	not reported, available in only 67% (8/12) patients	IV/10	emergency tracheostomy 10 days after distractor removal in syndromic patient
Andrews BT et al.[85]	internal nonresorbable	29% syndromic, exclude central sleep apnea, severe GER, or concomitant airway anomalies	Prevented: 97.26% (71/73)	N/A	dental injuries (6%), neck scarring (8%), wound infection (5%), temporomandibular joint ankyloses (10%)	64.8 months	IV/12	1 subject required tracheostomy for unrecognized central sleep apnea, 1 subject had emergent tracheostomy for loss of endotracheal tube on transport after surgery
Bangiyev JN et al.[88]	external	Syndrome not specified, 52% laryngomalacia, 24% tracheomalacia, 8% bronchomalacia	N/A	73.1% demonstrated severe pre-MDO sleep apnea (AHI>10); AHI: preoperative 30.3, postoperative 8.7 ($p<0.001$)	Not reported	at least 1 year	IV/12	
Breugem C et al.[89]	internal resorbable	17% syndromic, failed conservative therapy, exclude other airway obstruction	Prevented: 100.00% (11/11)	N/A	18% (1 cellulitis, 1 lost pin during consolidation)	not reported	IV/10	
Burstein FD et al.[80]	internal resorbable	25% syndromic, require supplemental O2, CPAP, intubation	Prevented: 100.00% (14/14); Decannulated: 83.33% (5/6)	preoperative RDI 15.34; postoperative RDI 1.11	28.5% (4 pin site infections)	24 months	IV/12	
Cascone P et al.[90]	external	7% syndromic, respiratory crisis at birth and endotracheal intubation	N/A	AHI: preoperative 80.1, postoperative 2.1	14% (4 loss of external pin during consolidation)	1 month	IV/12	
Chigurupati R et al. 91	internal nonresorbable	40% syndromic, fail conservative therapy	Prevented: 100.00% (2/2) Decannulated: 66.67% (2/3)	N/A	25% (pin site infection requiring early removal)	not reported	IV/10	
Ching JA et al.[83]	external	8% syndromic, no central apnea, laryngomalacia or tracheal stenosis	Prevented: 93.94% (31/33)	N/A	30% (4 pin site infection, 6 operative replacement of pin or device)	range 6 months to 10 years, mean 77.4 months	IV/12	1 failed due to laryngomalacia diagnosed postoperatively, previously not visualized
Denny A, Amm C[59]	external	63% syndromic, no secondary sites of airway obstruction	Prevented: 100.00% (11/11)	obtained in 7 patients, all previously severely impaired, normal 1 week to 1 month after operation	None	60 months	IV/12	1 patient previously failed TLA

MDO article	Type of Distractor	Patient selection/surgical indication	Tracheostomy prevented /Decannulation	OSA	Perioperative Complications	Follow up length	Level of Evidence/ MINORS	Notes
Denny A, Kalantarian B[58]	external	Syndromic status not specified, excluded patients with lower airway anomalies	Prevented: 100% (5/5)	all discharged home on apnea monitors, discontinued use after 90 days due to no apneic events	25% (1 device failure needing replacement)	range 9–22 months	IV/12	
Denny AD, Talisman R, Hanson PR et al. [29]	external	40% syndromic, fail conservative therapy	Decannulated: 66.67% (2/3)	10 subjects with predistraction apnea, none had triggering of apnea monitor 3 months post distraction	10% (1 pin displacement)	at least 6 months	IV/12	1 subject with Treacher Collins failed to distract because of premature consolidation and continued to require cannula
Flores RL, Greathouse ST, Costa M et al.[79]	internal nonresorbable	31% syndromic, include laryngomalacia, no central apnea, no other secondary airway anomaly	Prevented: 91.36% (74/81), 1 death due to apneic disease	8.11% (6) had AHI >20 postoperatively	Not reported	at least 1 year	IV/12	overlapping cohort with Tahiri et al, Murage et al., Tholpady et al., Flores RL, Tholpady SS, Sati S et al
Genecov DG et al.[66]	external and internal	4% syndromic, exclude laryngomalacia, tracheomalacia, subglottic/supraglottic stenosis, neurologic impairment, untreated GERD with vocal cord/epiglottic edema	Prevented: 96.15% (25/26) Decannulated: 92.00% (38/42)	preoperative RDI 35–50 to postoperative RDI 5–15 in 65 patients; in 2 patients RDI remained above 35 postoperatively	13% (9 pin site infections) 13% (9 device failure), 9% (6 asymmetric movement of depressor anguli oris muscle)	not reported	IV/10	2 failed decannulations due to previously undiagnosed swallowing abnormalities and inability to handle secretions
Goldstein JA et al.[77]	internal nonresorbable	46% syndromic, fail conservative therapy	Prevented: 87.50% (21/24) Decannulated: 100.00% (2/2)	AHI: preoperative PSGs in 20 subjects, 39.3±22.0; postoperative PSGs in 14 subjects, 3.0±1.5 ($p < 0.0001$)	4%(1 transient facial nerve paralysis), 8% (2 pin site infections), 4%(1 major submental abscess)	not reported	IV/10	1 patient underwent TLA after MDO
Gozu A et al.[92]	external	25% syndromic, fail conservative therapy	Prevented: 100% (4/4)	N/A	Not reported	44.4 months	IV/12	
Hammoudeh J et al. [93]	internal nonresorbable	25% syndromic, no lower airway anomaly	Prevented: 96.55% (28/29), 1 patient death	8 subjects requiring preoperative intubation: AHI preoperative 39.7, postoperative 3.13; remaining 20 subjects: AHI preoperative 39.7, postoperative 5.8.	14% (1 device failure, 1 transient facial nerve palsy, 1 device exposure, 1 pin site infection)	18.7 months	IV/12	1 tracheostomy in patient with interstitial lung disease and central apnea, 1 patient expired soon after surgery, (had multiple comorbidities)
Hong P et al.[65]	internal nonresorbable	67% syndromic, all had GERD, no central apnea, no lower airway anomaly	Prevented: 100.00% (6/6)	N/A	33% (2 local erythema)	18 months	IV/12	
Izadi K et al.[94]	internal nonresorbable	47% syndromic, central apnea, severe reflux, other airway lesions	Prevented: 93.33% (14/15)	N/A	None reported	14 months	IV/12	

MDO article	Type of Distractor	Patient selection/surgical indication	Tracheostomy prevented /Decannulation	OSA	Perioperative Complications	Follow up length	Level of Evidence/ MINORS	Notes
Konas E et al.[95]	internal nonresorbable	8% syndromic, fail conservative therapy	Prevented: 100.00% (13/13)	N/A	30% (1 facial nerve palsy, 1 reintubation, 1extension rod fracture, 2 pin site infections)	at least 1 year	IV/12	
Lam DJ et al.[39]	primarily external, few had internal	56% syndromic, fail conservative therapy	Prevented: 83.61% (51/61) Decannulation: 67.74% (42/62)	N/A	27% (14 premature consolidation, 9 open bite deformity, 5 TMJ ankylosis)	30 months	IV/12	poorer odds of success associated with craniofacial microsomia or Goldenhar syndrome (OR, 0.07 [95% CI, 0.009–0.52])
Lin SY et al.[96]	internal nonresorbable	None syndromic, all have OSA	Prevented: 100.00% (4/4) Decannulated: 100.00% (1/1)	3/5 had resolution of airway obstruction documented by PSG, AHI<1.5 and no snoring, 4th child had primary snoring without apnea, 5th child severe OSA with AHI 20.2	Not reported	median 47.5 months	IV/12	
Mahrous MA et al.[47]	internal nonresorbable	None syndromic, all had severe OSA	Decannulated: 100.00% (4/4)	preoperative RDI 14 to postoperative RDI 1	36% (incomplete osteotomy on one side, facial nerve palsy, pin site infection)	range 12–24 months	IV/12	
Mandell DL et al.[44]	external and internal	50% syndromic, exclude secondary airway anomaly, central sleep apnea, severe GERD	Prevented: 100.00% (8/8) Decannulated: 16.67% (2/12)	85% (5) had improvement of OSA, 3/6 had postoperative sleep studies showing resolution	23% (3 premature consolidation, 2 cheek abscess, 1 lip erosion, 1 cellulitis, 1 facial nerve palsy, 2 TMJ ankylosis)	13 months	IV/12	
Meyer AC et al.[97]	external	33% syndromic, fail conservative therapy	Prevented: 100.00% (18/18)	N/A	Not reported	79 months	IV/12	overlapping cohort with Tibesar et al. and Scott AR et al.
Mingo KM et al.[41]	external	only isolated PRS or Stickler's, compared patients with or without preoperative CT	Prevented: 98.04% (50/51) Decannulated: 100.00% (1/1)	N/A	12% (2 fracture between 2 posterior pins 1 bilateral hardware loosening, 1 temporary reintubation, 2 pin site infections, 1 unilateral marginal mandibular nerve weakness)	range 1–8 years	III/12	
Mitsukawa N et al.[98]	internal nonresorbable	30% syndromic, fail conservative therapy, have OSA	Prevented: 100.00% (8/8) Decannulated: 100.00% (2/2)	AHI: preoperative range 9.6 to 18.8, postoperative range 0.6 to 3.6	10% (device dislodged and reoperation)	range 6 months to 4 years	IV/12	
Monasterio FO, Drucker M, Molina F et al.[28]	external	Syndrome not specified, fail conservative therapy	Decannulated: 100.00% (3/3)	preoperative apnea index 20.5, hypopnea index 7.4; all resolved postoperatively	13% (1 device dislodged, 1 abscess)	3 months	IV/10	
Monasterio FO, Molina F, Berlanga F et al.[45]	external	6% syndromic, fail conservative therapy	N/A	Preoperative apnea index 18.3;	33% (5 pin site infection, 1 incomplete corticotomy requiring reoperation)	not reported	IV/8	

MDO article	Type of Distractor	Patient selection/surgical indication	Tracheostomy prevented /Decannulation	OSA	Perioperative Complications	Follow up length	Level of Evidence/ MINORS	Notes
				preoperative hypopnea index 8.5; both disappeared postoperatively				
Morovic CG, Monasterio L[99]	internal nonresorbable	29% syndromic, exclude central apnea and other airway anomalies	Prevented: 100% (5/5) Decannulated: 100.00% (2/2)	postoperative apnea index range 8 to 18	29% (2 pin extrusions)	range 1 to 45 months	IV/14	Prospective study
Mudd PA et al.[53]	internal nonresorbable	29% syndromic, fail conservative therapy	Prevented: 100.00% (24/24)	N/A	42% (1 facial paralysis due to compression of posteriorly displaced distraction arm, 2 facial nerve injury, 1 marginal mandibular weakness, 6 cellulitis)	28.8 months	IV/12	
Murage KP et al.[55]	internal nonresorbable	22% syndromic, fail conservative therapy	Prevented: 92.00% (46/50)	AHI: preoperative 37.8±25.6, postoperative 6.5±8.03 (p<0.05)	Device failure (2%), surgical site infection (22%), transient facial nerve palsy (2%).	37 months	IV/12	3 required repeat MDO
Rachmiel A, Emodi O, Rachmiel D et al.[100]	internal nonresorbable	Syndrome not specified, exclude tracheomalacia	Prevented: 100.00% (11/11) Decannulated: 100.00% (7/7)	Apnea index: preoperative >20, postoperative <2	5% pin site infection, 11% damage to mandibular branch of facial nerve	12 months	IV/12	
Rachmiel A, Srouji S, Emodi O et al.[46]	external	72% syndromic, severe OSA and tracheostomy	Decannulated: 100.00% (11/11)	Preoperative RDI>25, postoperative <2	Not reported	not reported	IV/8	
Scott AR et al.[101]	external	26% syndromic, fail conservative therapy	Prevented: 100.00% (17/17) Decannulated: 50.00% (1/2)	N/A	5% (1 anterior open-bite deformity), 21% (4 long-term tooth loss/malformation), 16% (injury to marginal mandibular branch of facial nerve), 16% (3 hypertrophic scars), 5% (1 repeat MDO)	67.2 months	IV/12	overlapping cohort with Tibesar et al. and Meyer et al.
Senders CW et al.[102]	external	23% syndromic, fail conservative therapy	Prevented: 90.91% (10/11) Decannulated: 100.00% (2/2)	N/A	15% (2 pin site infection) 8% (1 greenstick fracture), 8% (1 loose pins), 8% (1 mandibular branch of facial nerve palsy)	32.7 months	IV/12	subject requiring tracheostomy later diagnosed with central hypoventilatory syndrome
Shen W et al.[103]	internal nonresorbable	Syndrome not specified, fail conservative therapy	Prevented: 100.00% (6/6)	N/A	None reported	6 months	IV/10	
Schoemann MB et al.[104]	internal resorbable	18% syndromic, fail conservative therapy	Prevented: 100.00% (18/18)	N/A	5% (1 pin site infection)	not reported	IV/8	immediate 5mm at time of distractor placement
Sidman Jd et al.[105]	external	Isolated PRS only, fail conservative therapy	Prevented: 100.00% (2/2)	N/A	N/A	more than 3 years	IV/13	
Tahiri Y et al.[106]	internal nonresorbable	34% syndromic, exclude central apnea, include laryngomalacia	Prevented: 93.82% (76/81) Decannulated: 63.64% (7/11)	AHI: preoperative, 41.5; 1 month postoperative, 12.1; 1 year postoperative, 5.8	9% surgical site infection, 7.5% ventilator-associated pneumonia, 1.2% hematoma, 1.2% facial nerve neurapraxia	21.6 months	III/12	all infants <4kg, overlapping cohort with Flores RL, Greathouse ST, Costa M et al, Tholpady et al, and Murage et al.
Tholpady SS et al.[86]	internal nonresorbable	18% syndromic, all patients had laryngomalacia	Prevented: 100% (10/10) Decannulated: 100.00% (1/1)	AHI: preoperative	36% surgical site infection	28 months	IV/12	overlapping cohort with Flores RL, Greathouse

MDO article	Type of Distractor	Patient selection/surgical indication	Tracheostomy prevented /Decannulation	OSA	Perioperative Complications	Follow up length	Level of Evidence/ MINORS	Notes
				46.1±31.8, postoperative 4.1±3.0 (p = 0.002)				ST, Costa M et al, Murage et al, and Flores RL, Tholapdy SS, Sati et al.
Tibesar RJ et al.[54]	external	Syndrome percent not specified, fail conservative therapy	Prevented: 100.00% (32/32)	N/A	28% (9 open bite deformity), 16% (5 tooth malformation/ loss), 9% (3 facial nerve injury)	91.2 months	IV/12	overlapping cohort with Meyer AC et al. and Scott AR et al.
Wittenborn W et al. 81	external and internal	18% syndromic, exclude other airway anomalies	Prevented: 82.35% (14/17)	10 patients with both preoperative and postoperative PSG had 55% improvement in episodes per hour	None reported	16.5 months	IV/12	1 internal MDO required tracheostomy 4 months after surgery. 2 external MDO failed extubations, both had previously undiagnosed tracheal stenosis and required tracheostomy. 1 underwent correction and subsequent internal MDO and successful decannulation.
Zellner EG et al.[84]	internal nonresorbable	Syndrome not specified, fail conservative therapy	Prevented: 95.00% (19/20)	AHI: preoperative 51.3, postoperative 5.5 (p<0.1)	10% (2 device infection), 10% (2 facial nerve injuries)	not reported	III/12	multicenter study, 1 patient needed tracheostomy for continued sleep apnea

TLA studies	Distractor Type	Patient selection/surgical indication	Tracheostomy prevented	OSA	Complications	Follow up length	Level of Evidence/ MINORS	Notes
Abramowicz S et al. 78	N/A	15% syndromic, fail conservative therapy	90.00% (18/20)	N/A	Not reported	not reported	IV/11	Odds of requiring tracheostomy 5 times greater if GILLS score >3; both subjects requiring tracheostomy were syndromic
Bijnen CL et al.[63]	N/A	41% syndromic, fail conservative therapy	90.00% (18/20)	5/8 patients showed improvement in PSG	23% (5 partial dehiscence), 30% (6 small abscesses)	12 months (range 1 to 9 years)	IV/12	
Bull MJ et al.[82]	N/A	43% syndromic, fail conservative therapy	85.71% (6/7)	N/A	Not reported	range 8 to 30 months	IV/12	subject had multiple congenital anomalies with tracheobronchomalacia (found subsequently), required tracheostomy and CPAP for prolonged ventilatory management
Caouette-Laberge L et al.[2]	N/A	46% syndromic, fail conservative therapy	81.82% (9/11)	N/A	4 subjects deceased	not reported	IV/10	
Evans AK et al.[7]	N/A	46% syndromic, fail conservative therapy	83.87% (26/31)	N/A	6% (2 wound dehiscence)	not reported	IV/12	overlapping cohort with Rogers et al., 3 subjects requiring tracheostomy due to GER
Fujii M et al.[107]	N/A	18% syndromic, fail conservative therapy	89.47% (17/19)	N/A	Not reported	at least 1 year	IV/12	2 subjects (1 with Treacher Collins)

MDO article	Type of Distractor	Patient selection/surgical indication	Tracheostomy prevented /Decannulation	OSA	Perioperative Complications	Follow up length	Level of Evidence/ MINORS	Notes
								required tracheostomy at age 2 months; 1 for prolonged swelling of tongue on POD20; 1 required tracheal intubation soon after TLA and tracheostomy performed on POD24
Hoffman W[48]	N/A	70% syndromic, fail conservative therapy	91.30% (21/23)	N/A	26% (6 wound dehiscence)	39.6 months	IV/12	both patients requiring tracheostomy were syndromic, 1 subject did not have preoperative bronchoscopy, requiring tracheostomy for laryngomalacia; 1 subject required tracheostomy after recurrent respiratory infections and pneumonia, decannulated 7.5 years after tracheostomy
Huang F et al.[52]	N/A	Syndrome status not specified, fail conservative therapy	Prevented: 91.67% (11/12) Decannulated: 50.00% (1/2)	N/A	29% (4 wound dehiscence)	not reported	IV/8	subject requiring tracheostomy had multiple brain infarctions
Kirschner RE et al. 34	N/A	50% syndromic, fail conservative therapy	79.31% (23/29)	no obstructive apnea seen after takedown	41.6% dehiscence rate if mucosal adhesion alone, none if muscular sutures	29 months	IV/12	5/6 subjects requiring tracheostomy were syndromic; due to wound dehiscence (2), persistent glossoptosis, laryngomalacia, laryngeal granulation tissue, failure of later elective intubation
Li HY et al.[87]	N/A	Syndrome not specified, fail conservative therapy, require endotracheal intubation	42.86% (3/7)	N/A	4 failures of TLA due to wound dehiscence	at least 1 year	IV/12	
Mann RJ et al.[108]	N/A	All isolated PRS, fail conservative therapy, exclude tracheomalacia	95.45% (21/22)	N/A	13.6% (3 stitch abscesses), 4.5% (1 tongue mucocele)	96 months	IV/12	subject requiring tracheostomy had wound dehiscence secondary to traumatic postoperative extubation, eventually requiring tracheostomy for subglottic stenosis
Resnick CM et al.[49]	N/A	72% syndromic, fail conservative therapy, rule out other airway anomalies	100.00% (18/18)	AHI: preoperative 15.1±4.3, postoperative 9.9±4.1 (p=0.307)	Not reported	range 1 to 31 months	IV/11	
Rogers GF et al.[36]	N/A	45% syndromic, exclude other airway anomalies	88.68% (47/53)	N/A	Not reported	not reported	IV/11	overlapping cohort with Evans AK et al.; all subjects requiring tracheostomy were syndromic, 2 subjects due to early partial dehiscence and failed extubation, 1 due to laryngeal web and failed

MDO article	Type of Distractor	Patient selection/surgical indication	Tracheostomy prevented /Decannulation	OSA	Perioperative Complications	Follow up length	Level of Evidence/ MINORS	Notes
								subsequent MDO, 2 due to repeated aspiration, 1 due to GER and failure to wean off vent
Schaefer RB et al.[35]	N/A	All isolated PRS, rule out other airway anomalies	88.89% (8/9)	N/A	Not reported	median 33 months	IV/12	isolated PRS subject requiring tracheostomy due to previously unrecognized tracheomalacia
Sedaghat AR et al.[50]	N/A	Syndrome not specified, fail conservative therapy	87.50% (7/8)	AHI: preoperative AHI 52.6, postoperative 34.5; 1 subject had increase in AHI after TLA	Not reported	range 5 to 32 days	IV/11	subject requiring tracheostomy also failed MDO, had persistent respiratory issues and hypotonia

Comparison studies	Type of Distractor	Patient selection/surgical indication	Tracheostomy prevented	OSA	Perioperative Complications	Follow up length	Level of Evidence/ MINORS	Notes
Flores RL, Tholpady SS, Sati S et al.[42]	internal nonresorbable	Nonsyndromic only, sleep study results indicating an AHI >20, significant CO2 retention, absence of other significant airway anomalies, absence of TMJ abnormality	**MDO:** 100.00% (24/24); **TLA:** 73.33% (11/15), no p value provided	Preoperative AHI: MDO 47 vs TLA 37.6 ($p < 0.05$). Postoperative AHI: at 1 month, MDO 10.9 vs TLA 21.6 ($p < 0.05$), at 1 year, MDO 2.5 vs TLA 22. ($p < 0.05$)	**MDO: 16.67%** (1 equipment failure, 3 infections); **TLA: 40%** (3 wound dehiscence, 3 scar contractures); **no p-value**	at least 1 year	III/18	Retrospective, historical comparison, MDO (2004 to 2009) TLA (1994 to 2004)
Khansa I et al.[43]	internal nonresorbable	32% syndromic, unsustainable weight gain without tube feeds, unstable airway with positioning alone, no lower airway anomalies, no central sleep apnea, surgeon and family preference for MDO vs TLA	**MDO:** 100.00% (10/10); **TLA:** 100.00% (8/8)	**Preoperative AHI:** MDO 27.7, TLA 15.2 (p=0.01); **Postoperative AHI:** MDO 1.5, TLA 2.8 (p=0.8); mean decrease MDO 94.6 vs TLA 81.6% (p=0.01); **Residual moderate OSA** (AHI>5) in 0% MDO, 12.5% TLA (p=0.4)	**MDO: 40%** (4 cellulitis); **TLA: 25%** (2 temporary reintubations); **p=0.6**	1 year	II/19	prospective comparison, MDO subjects had highest baseline AHI (27.7), TLA subjects had intermediate baseline AHI (15.2)
Papoff P et al.[56]	external	56% MDO patients syndromic, 0% TLA patients syndromic, fail positioning and non-invasive ventilation, physical signs of respiratory distress associated with obstructive apneas, prolonged feeding difficulties, no distal airway anomalies	MDO: 88.89% (8/9); TLA 100% (9/9), **no p value provided**	surgery completely resolved airway obstruction in 3/9 TLA subjects, 8/9 MDO subjects (p=0.050); residual respiratory distress (opisthotonus,	**MDO: 44%** (4 lost pin in consolidation requiring repositioning), normal dental development; **TLA: 22%** (2 wound adhesions requiring repeat TLA), **no p value**	until age of palatoplasty (22.7±4.1 months in TLA group, 9.2±1.3 months in MDO group)	III/16	Retrospective; historical comparison, MDO (2006–2014), TLA (before 2002–2005)

MDO article	Type of Distractor	Patient selection/surgical indication	Tracheostomy prevented /Decannulation	OSA	Perioperative Complications	Follow up length	Level of Evidence/ MINORS	Notes
				pectus excavatum, desaturations, need for prone positioning), diagnosed more commonly after TLA than after MDO (6/9 vs 1/9, P=0.050).				
Susarla SM et al.[57]	internal nonresorbable	29.5% syndromic, continued feeding abnormalities (aspiration, penetration, or inability to control secretions) excluded patients needing pretreatment gastrostomy (global hypotonia, severe developmental delay) and multilevel airway lesions needing tracheostomy	MDO: 100.00% (30/30); TLA: 96.77% (30/31), **no p value provided**	N/A	**MDO: 17%** (4 surgical-site infections, 1 temporary marginal mandibular nerve palsy); **TLA: 10%** (3 wound dehiscence); **p nonsignificant**	3–4 years	III/18	2-institution retrospective; historical comparison, MDO (2010–2013), TLA (before 2007–2009)

MDO, mandibular distraction osteogenesis; TLA, tongue-lip adhesion.; N/A, not applicable; OR, odds ratio; CI, confidence interval; OSA, obstructive sleep apnea; RDI, respiratory distress index; AHI, apnea-hypoapnea index; PSG, polysomnogram; CPAP, continuous positive airway pressure; GER, gastroesophageal reflux

Table 3.

Comparison of Feeding Outcomes

Article	Type of Distractor	Patient selection/surgical indication	Full oral feeds	Growth / Weight	Complications	Follow up length	Level of Evidence/ MINORS	Notes
MDO studies								
Al-Samkari HT et al.[60]	external	42% syndromic, failure of conservative management	66.67% (8/12)	average daily weight gain 16.5 g	Not reported	not reported, available in only 67% (8/12) patients	III/10	60% of syndromic PRS needed gastrostomy compared to 0% of isolated PRS
Breugem C et al.[89]	internal resorbable	17% syndromic, failed conservative therapy, exclude other airway obstruction	90.91% (10/11)	N/A	18% (1 cellulitis, 1 lost pin during consolidation)	not reported	IV/8	6 subjects achieved full oral feeds at discharge, 4 more subjects within 4 weeks of discharge
Denny A, Amm C[59]	external	63% syndromic, no secondary sites of airway obstruction	100.00% (11/11)	growth above 50th percentile in all patients, trend of average or above-average weight gain continues in 4 patients with longest 3–5 year follow up	None	60 months	IV/12	1 patient previously failed TLA; 54.5% full oral feeds at 1 month, 100% at 1 year
Denny A, Kalantarian B[58]	external	Syndromic status not specified, excluded patients with lower airway anomalies	N/A	all subjects met or exceeded average 500g per month weight gain	25% (1 device failure needing replacement)	range 9–22 months	IV/12	
Genecov DG et al.[66]	external and internal	4% syndromic, exclude laryngomalacia, tracheomalacia, subglottic/supraglottic stenosis, neurologic impairment, untreated GERD with vocal cord/epiglottic edema	91.04% (61/67)	N/A	13% (9 pin site infections) 13% (9 device failure), 9% (6 asymmetric movement of depressor anguli oris muscle)	not reported	IV/10	
Goldstein JA et al.[77]	internal nonresorbable	46% syndromic, fail conservative therapy	83.33% (20/24)	N/A	4%(1 transient facial nerve paralysis), 8% (2 pin site infections), 4%(1 major submental abscess)	not reported	IV/10	1 patient underwent TLA after MDO
Izadi K et al.[94]	internal nonresorbable	47% syndromic, central apnea, severe reflux, other airway lesions	91.67% (22/24)	N/A	None reported	14 months	IV/12	

Article	Type of Distractor	Patient selection/surgical indication	Full oral feeds	Growth / Weight	Complications	Follow up length	Level of Evidence/ MINORS	Notes
Hong P et al.[65]	internal nonresorbable	67% syndromic, all had GERD, no central apnea, no lower airway anomaly	100.00% (6/6)	N/A	33% (2 local erythema)	18 months	IV/12	
Konas E et al.[95]	internal nonresorbable	8% syndromic, fail conservative therapy	100.00% (13/13)	N/A	30% (1 facial nerve palsy, 1 reintubation, 1extension rod fracture, 2 pin site infections)	at least 1 year	IV/12	
Mandell DL et al.[44]	external and internal	50% syndromic, exclude secondary airway anomaly, central sleep apnea, severe GERD	84.50% (7/8)	N/A	23% (3 premature consolidation, 2 cheek abscess, 1 lip erosion, 1 cellulitis, 1 facial nerve palsy, 2 TMJ ankylosis)	13 months	IV/12	1 patient required gastrostomy tube
Mingo KM et al.[41]	external	isolated PRS or Stickler's syndrome, compared patients with or without preoperative CT	86.54% (25/30)	N/A	12% (2 fracture between 2 posterior pins 1 bilateral hardware loosening, 1 temporary reintubation, 2 pin site infections, 1 unilateral marginal mandibular nerve weakness)	range 1–8 years	IV/12	
Mudd PA et al.[53]	internal nonresorbable	29% syndromic, fail conservative therapy	91.67% (22/24)	weight percentile: 25th after birth, significantly lower than norm during 3 month postoperative period, then steady growth curve through 1 year of age, most remain below 50th	42% (1 facial paralysis due to compression of posteriorly displaced distraction arm, 2 facial nerve injury, 1 marginal mandibular weakness, 6 cellulitis)	28.8 months	IV/12	50% discharged full oral feeds, 4 pts required gastrostomy tube, 3 were placed prior to MDO, 1 within 1 month of MDO for failure to thrive, at latest follow up 2 subjects continue to require supplemental feeding by gastrostomy tube (have associated neurologic sequelae of associated syndrome)
Scott AR et al.[101]	external	26% syndromic, fail conservative therapy	84.21% (16/19)	N/A	5% (1 anterior open-bite deformity), 21% (4 long-term tooth loss/ malformation), 16% (injury to marginal mandibular branch of facial nerve),	67.2 months	IV/12	overlapping cohort with Tibesar et al. and Meyer et al.

Article	Type of Distractor	Patient selection/surgical indication	Full oral feeds	Growth / Weight	Complications	Follow up length	Level of Evidence/ MINORS	Notes
					16% (3 hypertrophic scars), 5% (1 repeat MDO)			
Spring MA, Mount DL[61]	internal nonresorbable	90% syndromic, fail conservative therapy	70.00% (7/10)	7/10 subjects showed early decline in growth rate following distraction, persistent to 12 months; other 3 subjects had steady growth rate percentile	20% (2 mandibular nerve weakness) 30% (3 early limited mandibular range of motion), 10% (incisional scar hypertrophy)	range 12–28 months	IV/12	2 subjects required gastric tube, 1 gastric gavage; statistically significant relationship between younger age of subject and growth percentile rank decline
Tibesar RJ et al.[54]	external	Syndrome percent not specified, fail conservative therapy	78.13% (25/32)	N/A	28% (9 open bite deformity), 16% (5 tooth malformation/ loss), 9% (3 facial nerve injury)	91.2 months	IV/12	overlapping cohort with Meyer AC et al. and Scott AR et al.

TLA *studies*	Distractor Type	Patient selection/surgical indication	Full oral feeds	Growth / Weight	Complications	Follow up length	Level of Evidence	Notes
Bijnen CL et al.[63]	N/A	41% syndromic, fail conservative therapy	N/A	Catch up growth in 10 subjects, other 11 subjects remained on same growth percentile	23% (5 partial dehiscence), 30% (6 small abscesses)	12 months (range 1 to 9 years)	IV/12	
Caouette-Laberge L et al.[2]	N/A	46% syndromic, fail conservative therapy	100.00% (11/11)	N/A	4 subjects deceased	not reported	IV/10	

TLA Article	Type of Distractor	Patient selection/surgical indication	Full oral feeds	Growth / Weight	Complications	Follow up length	Level of Evidence	Notes
Cozzi F et al.[62]	N/A	Proportion syndromic not specified, fail conservative therapy	N/A	Body weight percentile: preoperative 9.7±2.6, postoperative 17.5± 4.6 (p>0.05); Weight velocity: preoperative 19.1±4.9, postoperative 74.2± 4.7 (p< 0.001)	19% (9 adhesion dehiscence)	68.4 months	IV/12	
Cruz MJ et al.[109]	N/A	17% syndromic, fail conservative therapy	83.33% (10/12)	N/A	17% (2 postoperative desaturations and bradycardia), 8% (1 chin abscess), 50% (6 dehiscences)	minimum 6 months	IV/12	

Article	Type of Distractor	Patient selection/surgical indication	Full oral feeds	Growth / Weight	Complications	Follow up length	Level of Evidence/ MINORS	Notes
Denny AD, Amm CA, Schaefer RB[51]	N/A	64% syndromic, fail conservative therapy	45.45% (5/11)	N/A	27% (3 wound dehiscence, 2 of which require repeat TLA)	94.8 months	IV/12	subjects gastrostomy dependent for more than a year
Hoffman W.[48]	N/A	70% syndromic, fail conservative therapy	65.22% (15/23)	N/A	26% (6 wound dehiscence)	39.6 months	IV/8	10 subjects weaned to oral feeds by day 21, 4 subjects discharged with gastrostomy tube, 9 with nasogastric tubes (5 converted to PO, 1 lost to follow up, and 3 required gastrostomy due to myopathy, aspiration on swallow study and severe oral aversion)
Huang F et al.[52]	N/A	Syndrome status not specified, fail conservative therapy	78.57% (11/14)	N/A	29% (4 wound dehiscence)	not reported	IV/10	
Kirschner RE et al.[34]	N/A	50% syndromic, fail conservative therapy	62.07% (18/29)	N/A	41.6% dehiscence rate if mucosal adhesion alone, none if muscular sutures	29 months	IV/12	
Schaefer RB et al.[35]	N/A	All isolated PRS, rule out other airway anomalies	100.00% (9/9)	N/A	Not reported	median 33 months	IV/12	those requiring nasogastric feeding in infancy discontinued by age 8 weeks

Comparison studies	Type of Distractor	Patient selection/surgical indication	Full oral feeds	Growth / Weight	Complications	Follow up length	Level of Evidence	Notes
Khansa I et al.[43]	internal nonresorbable	32% syndromic,unsustainable weight gain without tube feeds, unstable airway with positioning alone, no lower airway anomalies, no central sleep apnea, surgeon and family preference for MDO vs TLA	**MDO:** 90.00% (9/10), **TLA:** 87.50% (7/8), p=0.6	Similar baseline weight percentiles; change at 2–3 months: MDO −3.8% vs TLA −10.5%; at 10–12 month: MDO 19.5% vs TLA 19.4%; at 16–20 months: MDO 25.3% vs TLA 17.9%; failure to thrive duration: MDO 150 vs 162 days; all p > 0.5	**MDO: 40%** (4 cellulitis); **TLA: 25%** (2 temporary reintubations); p=0.6	1 year	II/19	prospective comparison, more MDO subjects required non-oral feeds at baseline (70 vs 37.5, p = 0.3); both patients requiring gastrostomy were syndromic

Article	Type of Distractor	Patient selection/surgical indication	Full oral feeds	Growth / Weight	Complications	Follow up length	Level of Evidence/ MINORS	Notes
Papoff P et al.[56]	external	56% MDO patients syndromic, 0% TLA patients syndromic,fail positioning and non-invasive ventilation, physical signs of respiratory distress associated with obstructive apneas, prolonged feeding difficulties, no distal airway anomalies	**MDO:** 100.00% (9/9), **TLA:** 100.00% (9/9)	N/A	**MDO: 44%** (4 lost pin in consolidation requiring repositioning), normal dental development; **TLA: 22%** (2 wound adhesions requiring repeat TLA), no p value	until age of palatoplasty (22.7±4.1 months in TLA group, 9.2±1.3 months in MDO group)	III/16	retrospective comparison; 1 patient with velocardiofacial syndrome and neurologic impairment (hypotonia) underwent TLA after MDO failed and received trach; Infants resumed oral feeding sooner after MDO than after TLA (mean days after surgery to full oral feeds 44±24 vs 217±134, (p<0.003).
Susarla SM et al.[57]	internal nonresorbable	29.5% syndromic, continued feeding abnormalities (aspiration, penetration, or inability to control secretions) excluded patients needing pretreatment gastrostomy (global hypotonia, severe developmental delay) and multilevel airway lesions needing tracheostomy	**MDO:** 83.33% (25/30), **TLA:** 15/31 (48.39%), p=0.009	N/A	**MDO: 17%** (4 surgical-site infections, 1 temporary marginal mandibular nerve palsy); **TLA: 10%** (3 wound dehiscence); p nonsignificant	3–4 years	III/18	2-institution retrospective; historical comparison, MDO (2010–2013), TLA (before 2007–2009)

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Table 4.

Summary of TLA and MDO Outcomes

Intervention	TLA	MDO
Avoidance of tracheostomy	89.47% (289/323) *	95.00% (657/693) *
Decannulation	N/A	80.00% (165/206)
Full Oral Feeds at latest follow up	70.06% (110/157)	87.30% (323/370)
Reported rates of reoperation	45% (5/11)[51], 29% (4/14)[52], 27% (3/11)[2], 22% (2/9)[56]	6% (3/50)[55], 5% (1/19)[54], 4% (1/23)[53]

*Five MDO studies, one TLA study and the MDO cohort of a comparison study were excluded from aggregation of results due to overlapping cohort of patients

MDO, mandibular distraction osteogenesis; TLA, tongue-lip adhesion.; N/A, not applicable.

Table 5.

Summary of Comparison Studies

Study	Design	Indications	Type of Distractor	Tracheostomy Prevented	Full oral feeds	Complications	Follow up length	Level of Evidence/MINORS
Flores RL, Tholpady SS, Sati S et al.[42]	Retrospective, historical comparison, MDO (2004 to 2009) TLA (1994 to 2004), nonsyndromic only, age<6 months,	sleep study results indicating an AHI >20, significant CO2 retention, absence of other significant airway anomalies, absence of TMJ abnormality	internal nonresorbable	**MDO:** 100.00% (24/24)**; TLA:** 73.33% (11/15), **no p value provided**	N/A	**MDO: 16.67%** (1 equipment failure, 3 infections); **TLA: 40%** (3 wound dehiscence, 3 scar contractures); **no p-value**	at least 1 year	III/18
Khansa et al.[43]	Prospective, 32% syndromic, nonrandomized, MDO subjects had higher baseline AHI (27.7) vs TLA (15.2)	unsustainable weight gain without tube feeds, unstable airway with positioning alone, no lower airway anomalies, no central sleep apnea, surgeon and family preference for MDO vs TLA	internal nonresorbable	**MDO:** 100.00% (10/10)**; TLA:** 100.00% (8/8)	**MDO:** 90.00% (9/10)**, TLA:** 87.50% (7/8), p=0.6	**MDO: 40%** (4 cellulitis); **TLA: 25%** (2 temporary reintubations); p=0.6	1 year	II/19
Papoff et al.[56]	Retrospective; historical comparison, MDO (2006–2014), TLA (before 2002–2005), 56% MDO patients and 0% TLA patients syndromic, no distal airway anomalies	fail positioning and non-invasive ventilation, physical signs of respiratory distress associated with obstructive apneas, prolonged feeding difficulties	external	**MDO: 88.89% (8/9),** patient with veocardiofacial syndrome and neurologic impairment (hypotonia) underwent TLA after MDO failed then received tracheostomy; **TLA 100% (9/9), no p value,** 1 patient died (after TLA lysis, did not respond to resuscitation); **residual respiratory distress** diagnosed more commonly after TLA than after MDO (6/9 vs 1/9, P=0.050).	**MDO: 100.00% (9/9), TLA: 100.00% (9/9),** infants **resumed oral feeding** sooner after MDO than after TLA (44±24 vs 217±134 days, P<0.003).	**MDO: 44%** (4 lost pin in consolidation requiring repositioning), normal dental development; **TLA: 22%** (2 wound adhesions requiring repeat TLA), no p value	until age of palatoplasty (22.7±4.1 months in TLA group, 9.2±1.3 months in MDO group)	III/16

Study	Design	Indications	Type of Distractor	Tracheostomy Prevented	Full oral feeds	Complications	Follow up length	Level of Evidence/MINORS
Susarla et al.[57]	2-institution retrospective; historical comparison, MDO (2010–2013), TLA (before 2007–2009), groups statistically comparable in demographic and clinical factors, 29.5% syndromic, excluded patients needing pretreatment gastrostomy (global hypotonia, severe developmental delay) and multilevel airway lesions needing tracheostomy	fail conservative management, continued feeding abnormalities (aspiration, penetration, or inability to control secretions)	internal nonresorbable	**MDO:** 100.00% (30/30)**;** **TLA:** 96.77% (30/31)**, no p value, 1 death in TLA cohort**	**MDO:** 83.33% (25/30), **TLA:** 15/31 (48.39%), **p=0.009**	**MDO: 17%** (4 surgical-site infections, 1 temporary marginal mandibular nerve palsy); **TLA: 10%** (3 wound dehiscence); p nonsignificant	3–4 years	III/18

Table 6.

Comparison of TLA and MDO advantages and disadvantages

Intervention	Advantages	Disadvantages	Commonly cited complications
TLA	immediate relief of airway obstruction, short operating time, shorter length of stay and overall cost[6], minimal external scarring	reliance of "catch-up growth" of the mandible, dysphagia and early feeding difficulties[51], interference with development of pre-speech skills and sound production[110]	wound dehiscence, early release of retention sutures, tongue lacerations, injuries to Wharton's ducts
MDO	lengthens mandible and increases mandibular volume, lower cost compared to tracheostomy[111]	requires specialized training and equipment, parental compliance in turning device, second operation required for removal, gradual improvement of airway obstruction	hypertrophic scarring, device malfunction or infection, open bite deformity, dental damage, facial nerve injury