Airway management in Pierre Robin sequence: The Vancouver classification

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BACKGROUND: Pierre Robin Sequence (PRS) is a triad of micrognathia, glossoptosis and respiratory distress. There is no standard clinical classification used in the management of neonatal airway in PRS patients. The goal of our study was to review the presentation and management of PRS patients and formulate a clinical grading system and treatment algorithm.

METHODS: A 10-year retrospective review of all neonates diagnosed with PRS was performed after obtaining institutional ethics approval. Patients were identified using our cleft lip and palate database. Inclusion criteria were 2 out of the following 3 clinical features: glossoptosis, retrognathia or airway obstruction. We collected demographic data, clinical information (co-existing airway morbidity, maxillary-mandibular discrepancy (MMD), type of intervention employed, complications and outcomes (feeding, length of stay, airway status) during the first year of life.

RESULTS: 63 patients met our inclusion criteria. Of these, 55 (87%) had cleft palate and 17 (27%) were syndromic. 48 patients (76%) were managed by prone positioning. Of the 15 surgically-managed patients, the initial procedure was floor of mouth release (FMR) in 7, mandibular distraction osteogenesis (MDO) in 4, and tongue-lip adhesion (TLA) in 4. 5 patients with co-existing airway morbidity needed a second surgery; 2 had MDO and 3 tracheostomies (one was later decannulated). 7 (47%) of the surgically-managed patients required a gastrostomy tube.

CONCLUSION: At present there is no consensus on neonatal airway management in PRS infants. From our review of 63 PRS patients, we hereby propose a simple 4-point classification system and treatment algorithm, based on clinical features.

Key Words: *Pierre Robin sequence; Classification; Mandibular distraction osteogenesis; Tongue lip adhesion; Floor of mouth release; Respiratory distress*

La gestion des voies aériennes dans Pierre Robin Séquence: La classification de Vancouver

CONTEXTE: La séquence de Pierre Robin (PRS) est une triade de micrognathie, de glossoptosis et de détresse respiratoire. Il n'existe pas de classification clinique standard utilisée dans la prise en charge des voies respiratoires néonatales chez les patients PRS. L'objectif de notre étude était d'examiner la présentation et la gestion des patients PRS et de formuler un système de classement clinique et un algorithme de traitement.

Méthodes: Un examen rétrospectif sur 10 ans de tous les nouveau-nés diagnostiqués avec PRS a été réalisé après avoir obtenu l'approbation éthique de l'établissement. Les patients ont été identifiés à l'aide de notre base de données sur les lèvres et les palais. Les critères d'inclusion étaient 2 des 3 caractéristiques cliniques suivantes: glossoptose, rétrognathie ou obstruction des voies aériennes. Nous avons recueilli des données démographiques, des données cliniques (morbidité coexistante des voies aériennes, écart maxillaire-mandibulaire (MMD), type d'intervention employée, complications et résultats (alimentation, durée du séjour, état des voies aériennes) au cours de la première année de vie.

RÉSULTATS: 63 patients répondent à nos critères d'inclusion. Parmi ceux-ci, 55 (87%) avaient une fente palatine et 17 (27%) étaient syndromiques. 48 patients (76%) ont été pris en charge par un positionnement ventral. Sur les 15 patients chirurgicaux, la procédure initiale était le stade de libération de la bouche (FMR) dans 7, l'ostéogenèse de la distraction mandibulaire (MDO) en 4 et l'adhérence de la langue et de la lèvre (TLA) chez 4, 5 patients présentant une morbidité coexistante des voies aériennes Besoin d'une deuxième intervention chirurgicale; 2 avaient MDO et 3 trachéostomies (un a été décannulé plus tard). 7 (47%) des patients chirurgicaux ont besoin d'un tube de gastrostomie.

CONCLUSION: À l'heure actuelle, il n'y a pas de consensus sur la prise en charge néonatale des voies respiratoires chez les nourrissons PRS. Dans notre revue de 63 patients PRS, nous proposons un système simple de classification en 4 points et un algorithme de traitement basé sur des caractéristiques cliniques.

Pierre Robin reported a series of neonates with a triad of micrognathia, glossoptosis and respiratory distress in 1923 (1). The association of cleft palate was added to the sequence in 1934 (2). Despite earlier descriptions by others, the eponym of Pierre Robin sequence has stood the test of time, although confusion in nomenclature remains (3). Regardless, these infants present with respiratory distress, manifested by stridor and desaturations, as well as feeding difficulties and failure to thrive. To add to the confusion, the spectrum of symptom severity varies significantly, from problems with feeding and weight gain, to airway obstruction and sleeping difficulties. This results in a lack of consistency in the management of these patients.

At present, no classification system exists to define PRS patients. A universal clinical classification system would facilitate better communication between individuals on the multidisciplinary team that are involved in the care of these patients. Furthermore, a classification system would allow guidance in management and comparison of clinical outcomes between institutions.

Upon review of the literature, there is one retrospective study that divides PRS infants into three groups according to severity of respiratory symptoms and the mode of feeding. This study was published in 1994, prior to the use of mandibular distraction osteogenesis in this population and does not separate out the different surgical approaches used (4). There is a more recent study describing a grading system based on clinical nursing protocol, focused on positioning the infant, need for nasopharyngeal airway and nasogastric feeding (5). Again, this paper does not take into account the need for surgical intervention, nor the various surgical approaches used to manage respiratory distress in these patients.

Current management of PRS infants has been recently reviewed (6) and can be divided into non-surgical techniques and surgical procedures. Non-surgical techniques include prone or lateral positioning, nasopharyngeal or endotracheal airway and CPAP. Surgical interventions currently used for the management of airway obstruction are floor of mouth release (FMR), tongue-lip adhesion (TLA), mandibular distraction osteogenesis (MDO) and tracheostomy.

Neonates who are managed successfully with non-surgical methods have a milder anatomical problem, which is resolved after a period of 'partial catch-up' mandibular growth. The latter was demonstrated in a retrospective longitudinal cephalometric study comparing PRS vs. isolated cleft palate children vs. normal controls (7). However, clearly only a subset of PRS patients exhibit sufficient 'catch up' growth to avoid the need for surgery (8).

Each surgical intervention comes with inherent risks and benefits.

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At present, it seems that the surgical approach employed by different institutions is dependent on the individual surgeon's preference rather than a logical algorithm (9). Since Denny et al published the use of MDO for the management of Pierre Robin infants (10), this has become increasingly popular in many centres. A recent survey of members of the American Cleft Palate-Craniofacial Association reported that 48% of the 87 respondents preferred the use of MDO for airway management in PRS infants (11). In another recent study, the senior surgeon switched from TLA to MDO and reported superior outcome in terms of oxygen saturation, apnea-hypopnea index and need for tracheostomy (12). Some authors reserve MDO for patients with failed TLA or FMR (13-15).

Due to the relatively low incidence of PRS, coupled by the fact that only a minority of patients require surgical intervention, there is a lack of prospective comparison studies between MDO, TLA and FMR. The ultimate goal in these infants is to perform a single operation that will successfully manage the airway, improve the ability to feed and avoid the morbidity of tracheostomy. While TLA and FMR have shown to be successful in a subset of infants, it is clear that this is not the optimal procedure in all patients and doomed to fail in some. This latter group would be better served with MDO as their initial and only procedure. It would therefore be helpful to devise an algorithm to categorize these various groups of infants to predict which are most likely to succeed with the different surgical approaches.

The goals of this study were to review our experience with PRS infants managed over a 10-year period and devise a universal classification system, based on clinical severity. Using this classification system, we propose a management algorithm to minimize the number of procedures needed for successful airway management in PRS patients.

METHODS

Following approval from the University of British Columbia Children's and Women's Research Ethics Board (# H14-00353), we performed a 10-year retrospective chart review of all patients with Pierre Robin Sequence, seen between 2004 and 2013. Patients were identified using the hospital discharge database, as well as the cleft palate clinic database. All patients (syndromic or non-syndromic) identified as having two of the following features were included: glossoptosis, micrognathia or airway obstruction. Demographic data collected included: gender, gestational age, associated syndromes, presence of cleft palate, symptoms of failure to thrive, airway obstruction, desaturations, feeding difficulties, nonsurgical and surgical airway approaches, feeding management (within the first year of life), age at surgical intervention and outcomes of management (symptom recurrence, length of hospital stay and the need for further intervention). All data was analyzed using descriptive statistics.

Subjects were identified as having airway obstruction if any of the following signs of airway obstruction were mentioned in their chart: stridor, noisy breathing, tracheal tug, intercostal retractions or cyanosis. Subjects were defined as having desaturations if any single recorded hospital oxygen saturation was less than 80 percent or if more than one hospital oxygen saturation was less than 90 percent. Our institution does not routinely perform polysomnography for these neonates to document apnea, but rely on oximetry to document desaturations. Subjects were identified as having difficulty feeding, poor weight gain/failure to thrive if feeding time exceeded 30 minutes, if patients had <30g/24hours weight gain or if the patient required nasogastric tube or gastrostomy tube feedings.

All patients that failed non-operative management underwent surgical treatment, after nasendoscopy and bronchoscopy, to exclude pre-existing airway pathology. Depending on each individual surgeon's preference, techniques performed were FMR, TLA and MDO. Tracheostomy was reserved for those patients with failed FMR and MDO.

RESULTS

A total of 63 patients were managed for PRS during our study period. 55 (87%) of these patients had cleft palate (Table 1). There were 30 females and 33 males. 17 patients were syndromic, with the most common syndrome being Stickler.

48 patients (76%) were managed successfully by non-surgical treatment (Figure 1). 15 patients required surgical management of the airway, with a mean age of 51 days at the time of surgery (ranging from 5–246 days). 10 (67%) required only a single procedure, but 5 (33%) ultimately needed a second surgery (Table 2). The initial surgical intervention was floor of mouth release (FMR) in 7 patients, tongue-lip adhesion (TLA) in 4 patients and mandibular distraction osteogenesis (MDO) in 4. In the 5 patients who needed multiple procedures, 3 ultimately required tracheostomies and 2 were managed successfully with a subsequent MDO. Of the patients managed surgically, 7 demonstrated feeding difficulties post-operatively and required a gastrostomy tube.

Successful single-procedure airway management was achieved in all 4 patients undergoing TLA, 3 of 7 for FMR and 3 out of 4 MDO. Of the 5 patients who had a failed FMR, 3 had co-existing airway morbidity (tracheomalacia, laryngomalacia and bronchomalacia). 2 of these patients subsequently did well with MDO and the other patient was managed with tracheostomy. The one patient with failed MDO, had laryngomalacia and two attempts at supra-glottoplasty by the otolaryngology service. That patient also had congenital VI and VII cranial nerve palsies.

DISCUSSION

Pierre Robin Sequence patients are a heterogeneous group of infants that present with a combination of respiratory distress and feeding difficulties. In keeping with the literature, we found that the majority of our patients (76%) could be managed successfully by non-surgical means such as prone positioning and nasopharyngeal airway (15,16). We found that the majority of patients (87%) had cleft palate. 14 of 15 (93%) of those infants requiring surgery had cleft palate compared to 87% of those managed without surgery. Overall, 27% of PRS patients in our series were syndromic. It has been suggested that infants with PRS as part of a syndrome had poorer outcomes than those with isolated PRS (17). This is consistent with the present series, as we found that the number of syndromic patients in the surgical group (7 out of 15, 47%) was higher than those managed without surgery (10 out of 48, 21%).

For those that require surgical intervention, the ultimate goal is to perform a single procedure for successful airway management and avoid tracheostomy. Procedures are broadly divided into tongue repositioning techniques (TLA and FMR) and mandible lengthening, namely MDO. Of the 15 patients who underwent surgery, all 4 (100%) patients who underwent TLA had successful surgery and did not require any further intervention (Figure 1 and Table 2). This is consistent with other reports of success rates of 71-89% for TLA and is frequently considered first-line surgical treatment of those failing non-operative management (12). It is important to note that none

TABLE 1
Primary diagnosis of patients managed non-surgically compared with surgically managed patients

	Non-Surgical	Surgical	Total
Number of patients	48	15	63
Female	25	5	30
Male	23	10	33
Cleft Palate			
Yes	41	14	55
No	7	1	8
Non-Syndromic	36	8	44
Syndromic	10	7	17
Stickler	4	5	9
Hemi-facial Microsomia	2	1	3
Oromandibular Limb	1	1	2
Hypoplasia	1	0	1
Craniosynostosis	1	0	1
Smith-Magenis	1	0	1
Genitopatellar			

TABLE 2
Demographics of surgically managed PRS patients

Subject Number	Gest. Age	CP	Syndrome	MMD	Ongoing O_2 de-saturations	Ongoing feeding difficulties	Airway pathology	Initial Sur-gery	Age at surgery (days)	2nd Surgery	LOS (days after surgery)	Feeding at 6 months post op	Complications
8	37	Yes	No	14mm	Yes	Yes	none	TLA	60	None	106	Bottle	None
16	37	Yes	Stickler	4 mm	Yes	Yes	none	TLA	51	None	16	Bottle	None
17	40	Yes	No	8-10mm	Yes	Yes	none	TLA	9	None	28	Bottle	None
74	36	Yes	Stickler	3 mm	Yes	Yes	none	TLA	25	None	11	Bottle	None
54	40	Yes	No	<10mm	Yes	Yes	none	FMR	246	Tracheostomy (decannulated)	57	G tube	Failed extubation and cardiac arrest (survived)
66	38	Yes	No	5 mm	Yes	Yes	none	FMR	9	None	80	G tube	None
60	39	Yes	Stickler	<10mm	Yes	Yes	tracheomalacia and tracheo-esophageal fistula	FMR	19	Tracheostomy (permanent)	29	G tube	Failed extubation
64	29	Yes	Stickler	<10 mm	Yes	Yes	none	FMR	109	None	13	Bottle	None
20	36	Yes	Genito-patellar	<10mm	Yes	Yes	laryngomalacia	FMR	43	MDO	24	G tube	None
40	38	Yes	Stickler	<10mm	Yes	Yes	bronchoma-lacia	FMR	5	MDO	52	Bottle	Right pneumothorax and failed extubation after FMR
46	37	Yes	No	>10mm	Yes	No	none	FMR	6	None	42	Bottle	None
1	37	Yes	No	14 mm	Yes	Yes	none	MDO	54	None	15	Bottle	None
4	40	Yes	No	1 mm	Yes	Yes	none	MDO	92	None	15	G tube	Developed Oral Aversion
34	39	Yes	No	>10 mm	Yes	Yes	tracheomalacia	MDO	29	None	154	G tube	Infection of device and cheek abscess and osteomyelitis
63	38	No	Oro-mandibular Limb Hypoplasia	12mm	Yes	Yes	laryngomalacia and tracheomalacia	MDO	9	Tracheostomy (permanent)	150	G tube	None

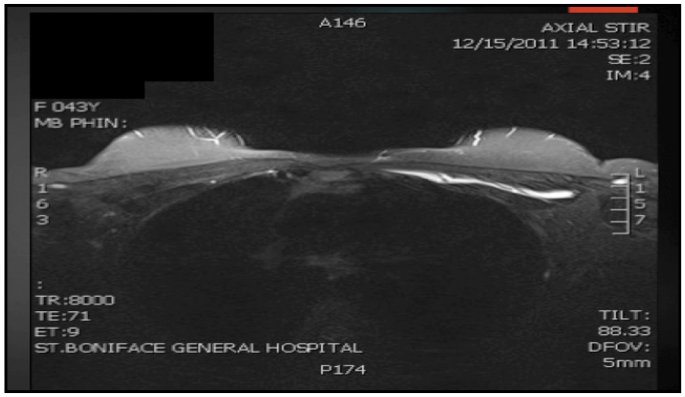

Figure 1) *Flowchart showing initial and secondary surgical procedures in surgically managed patients*

of these patients had co-existing airway pathology such as tracheomalacia or laryngomalacia.

Of our patients undergoing FMR, 3 out of 7 patients were successfully managed with FMR alone. Of the 4 that needed further surgical intervention, 3 patients had associated airway pathology, namely bronchomalacia, tracheomalacia and tracheo-esophageal fistula and larygnomalacia. Of these, 2 were managed successfully by MDO and the third patient was managed by tracheostomy, at a time prior to our institution routinely performing MDO. Our findings suggest that patients with associated airway pathology are unlikely to be managed successfully with FMR alone and should undergo MDO as the initial approach.

At our institute, MDO has been used since 2008 for patients with either a co-existing airway pathology or maxillo-mandibular discrepancy of ≥10mm. 3 of the patients who had MDO as the initial procedure and 2 of the patients who had MDO after failed FMR, were successful (83%) and did not require any further intervention. Only one patient, (subject 63), who underwent MDO as the initial procedure, at the age of 9 days, failed extubation and ultimately had a tracheostomy. This patient also had congenital VI and VIIth cranial nerve palsies, laryngomalacia and tracheomalacia. His post-operative course was complicated by cellulitis of the abdominal wall from the gastrostomy tube site.

There are inherent limitations to our study, due to several factors, including the retrospective design, the variability among the 4 surgeons involved, and the small sample size. The latter is partly a result of the low incidence of PRS and the high number of patients managed non-surgically. Nonetheless, our results show that the selection of patients for surgery and the chance of surgical success can be improved by correctly identifying and addressing the etiology of the airway obstruction on an individual basis. At present, there still appears to be a culture of employing the surgical technique based on surgeons' preference (9,11). We propose that each PRS patient poses a specific etiology, whether it is predominantly intrinsic airway pathology, glossoptosis, or mandibular hypoplasia, and this should be addressed surgically, instead of a 'one size fits all' philosophy.

Vancouver Classification for Airway Management of PRS

From our findings, we present a clinical classification system of Pierre Robin Sequence patients (Table 3). This 4-grade system, from Grade 0 to 3, is dependent on the presence and absence of key clinical features and can be used to determine methods of non-surgical and surgical management.

- Grade 0- Patients with the mildest presentation, have no co-existing airway pathology, MMD <10 mm with mild glossoptosis and no feeding difficulties. These patients respond to non-surgical management, such as prone positioning.
- Grade I– These patients have MMD <10mm, moderate or severe glossoptosis, ongoing feeding difficulties and NG dependency. They have no co-existing airway pathology and have ongoing desaturations, despite prone positioning. In these patients, we would recommend a tongue repositioning procedure, either TLA or FMR.
- Grade II– These patients have MMD≥10mm, with moderate or severe glossoptosis, ongoing desaturations with prone positioning, ongoing feeding difficulties, NG dependency, co-existing airway pathology and fail to respond to non-surgical means. These patients are unlikely to respond to soft tissue tongue-repositioning surgery, given the severity of mandibular hypoplasia, and should undergo MDO.
- Grade III– These patients have the severest pathology and if after failed MDO, should undergo tracheostomy.

TABLE 3
The Vancouver classification and treatment algorithm

Severity Grade	Clinical Features	Treatment
Grade 0	1. MMD <10mm 2. Mild glossoptosis 3. Maintains saturations with prone positioning 4. No feeding difficulties 5. No co-existing airway morbidity	Prone positioning or nasopharyngeal airway
Grade I	1. MMD <10mm 2. Moderate or severe glossoptosis 3. Ongoing desaturations with prone positioning 4. Ongoing feeding difficulties and NG dependent 5. No co-existing airway morbidity	Tongue lip adhesion or floor of mouth release
Grade II	1. MMD >10 mm 2. Moderate or severe glossoptosis 3. Ongoing desaturations with prone positioning 4. Ongoing feeding difficulties and NG dependent 5. Co-existing airway morbidity (laryngomalacia, tracheomalacia or bronchomalacia)	Mandibular distraction osteogenesis
Grade III	1. MMD >10 mm 2. Severe glossoptosis 3. Ongoing desaturations with prone positioning 4. Ongoing feeding difficulties and NG dependent 5. Co-existing airway morbidity (laryngomalacia, tracheomalacia or bronchomalacia) 6. +/- Failed MDO	Tracheostomy

CONCLUSION

There is currently no consensus in the surgical management of respiratory distress in patients with PRS that fail to respond to non-surgical management. Following a 10-year retrospective review of 63 PRS patients, we hereby propose a simple 4-point classification system, based on clinical features. Using our clinical grades we suggest a corresponding treatment algorithm to guide management.

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