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Nasopharyngeal Tube and Functional Treatment in Pierre Robin Sequence: A Tertiary Clinical Experience From 150 Cases

Fabian Blanc, MSc¹, Inge Harrewijn, MD², Claire Duflos, MD³, Frederica Maggiulli, MD⁴, and Guillaume Captier, MD, PhD^{5,6}

Abstract

Objectives: To describe the initial care practices for children with Pierre Robin sequence (PRS) and analyze the factors predicting the severity of the obstruction breathing disorders.

Design: A retrospective single-center study of 150 children with PRS.

Setting: Single tertiary care center, Regional Competence Center for the diagnosis and treatment of PRS.

Patients: A total of 150 children with PRS consecutively followed between 1986 and 2017. Group 1 comprises children without specific respiratory management; group 2, children requiring prone positioning to alleviate their respiratory distress symptoms; and group 3, children requiring nasopharyngeal airway tube (NT) or nonconservative surgical treatment.

Main outcome measures: Evolution and results of the initial treatment of PRS.

Results: Forty-two percent ($n = 63$) were attributed to group 1, 39% ($n = 50$) to group 2, and 19% ($n = 29$) to group 3. Preterm birth, birth weight, or associated congenital malformations were not significantly different between the groups. However, the age of exclusive oral feeding was significantly different: 1 day (quartiles: 0-3) for group 1; 11 days (quartiles: 1-28) for group 2; 39 days (quartiles: 19-111) for group 3 ($P < .0001$). Considering the NT, its use relieves the upper airway obstruction, assessed by a respiratory polygraphy, in 14 children.

Conclusions: Nasopharyngeal airway tube has become our major first-line treatment, avoiding more complex procedures in most of the cases. The achievement of exclusive oral feeding seems to be a good predictor of the severity of respiratory symptoms in PRS.

Keywords

Pierre Robin sequence, respiratory distress, nasopharyngeal tube, upper airway obstruction, cleft palate

Introduction

The Pierre Robin sequence (PRS) is a congenital malformative sequence and the micrognathia is the most accepted primary event. Pierre Robin sequence can be isolated or associated with other anomalies and in this case is a syndromic PRS. Although declining, mortality rate for children with PRS reaches up to 16.6% (Logies et al., 2018). The incidence of this pathology is 1/8500 live births in the occidental population (Vatlach et al., 2014). Since it is associated with glossoptosis, the abnormal posterior and vertical position of the tongue can lead to a U-shaped cleft palate (Robin, 1934; Hanson & Smith, 1975). These anatomical anomalies are responsible for breathing difficulties and suction-swallowing incoordination (Baudon et al.,

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2002), which can be worsen by brainstem dysfunction associated with PRS (Abadie et al., 2002). The consequences in terms of respiratory and feeding are widely variable between children with PRS (Breugem et al., 2016). In addition, the level of micrognathia and glossoptosis did not seem to predict the severity of symptoms of this condition (Morice et al., 2018).

There is no consensus for the initial care for these patients in the literature. Many children may be managed conservatively with only nursing instructions, but some others present severe life-threatening upper airway obstruction requiring specific urgent treatment. These treatments range from the use of a nasopharyngeal airway device (de Buys Roessingh et al., 2007), surgical interventions, including mandibular distraction (Rathé et al., 2015), to tracheostomy. The incidence of tracheostomy in PRS is reported to be up to 15% in a recent study (Evans et al., 2006), although most recent studies seem to demonstrate that nonsurgical management is preferred in PRS (Morice et al., 2018; Gómez et al., 2018a). The success in the use of nasopharyngeal airway device could explain this evolution (Maas & Poets, 2014; Drago Marquezini Salmen & Lazarini Marques, 2015). A prospective multicenter blind study attests to the high efficiency of the pre-epiglottic baton plate (PEBP; Poets et al., 2017).

In our tertiary center, the conservative nonsurgical attitude in the treatment of children with PRS was our first approach with a well-established use of the nasopharyngeal airway tube (NT) since 2001. The NT is less invasive, with low morbidities (de Buys Roessingh et al., 2007) and probably less expensive than surgical treatments or PEBP. Our hypothesis is that our protocol including correct use of NT is sufficient to relieve obstruction of the upper respiratory tract for most PRS children. Nasopharyngeal airway tube also makes it possible to feed the baby orally more quickly than with surgical treatments, ensuring oral stimulation and adequate weight gain.

The main objective of this retrospective study was to describe the evolution of the initial treatment of PRS in a single tertiary center. The secondary objectives were to present the results of the use of the NT and to analyze the predictive factors for the severity of respiratory obstruction.

Material and Methods

This is a retrospective single-center study concerning 150 children with PRS consecutively followed in the tertiary University Hospital, Regional Competence Center for the diagnosis and treatment of PRS (Filière maladie rare TêteCou), over a 30-year period (from 1986 to 2018). The local ethic committee of the university hospital was obtained (2019_IRB-MTP_10-08).

According to the data from the French National Institute of Statistics during this period, there were 874 640 live births in the region (Statistiques | Insee) (2020), resulting in an incidence of PRS of 1/6000 live births, concordant with the incidence found by Vatlach et al. (2014) in Germany, suggesting low follow-up loss rate.

These children were taken care of by our multidisciplinary team from neonatology, pediatrics, sleep medicine, pediatric plastic surgery, pediatric otolaryngology, orthodontics, speech therapy, and genetics.

Data from these patients were collected from birth to the age of 6 months of life.

Inclusion Criteria of Patients With PRS

Pierre Robin sequence associated with U-shaped cleft palate was chosen from the medical database neonates. The presence of a U-shaped cleft palate as a selection criterion was chosen because this definition was durably used to diagnose PRS in our center during the 30-year period of this study. The PRS without cleft palate was not included.

As reported in the clinical consensus report (Breugem et al., 2016), the characterization of the severity of the anatomical malformations (micrognathia and glossoptosis) varies between the teams, and no standard method exists to date. In practice, micrognathia was characterized by considering the maxillo-mandibular discrepancy clinically and glossoptosis was defined as a displacement of the tongue posteriorly and vertically, associated with a direct abnormal view of the sublingual crests at gentle mouth opening. Children with smaller than average jaws but no evidence of tongue base airway obstruction were not included as patients with PRS.

Clinical Data

Clinical data reported at birth were prenatal diagnosis of PRS, term, size, weight and circumference of the head at birth, and neonatal Apgar score (American Academy of Pediatrics Committee on Fetus and Newborn and American College of Obstetricians and Gynecologists Committee on Obstetric Practice, 2015).

Following a multidisciplinary assessment, including clinical genetics, PRS was classified after 6 months of life as: isolated PRS (iPRS), syndromic PRS (sPRS), or PRS associated with other clinical abnormalities that did not match diagnostic criteria for no known syndrome (aPRS).

Respiratory Treatment

Respiratory treatments required for our patients with PRS during their first 6 months of life were: (1) lying or lateral positioning, (2) use of an NT, or (3) surgical treatment.

- (1) Prone or lateral positioning: The children were nursed in the supine or lateral position with linen under the belly or the lateral back with cardiac and respiratory monitoring during hospitalization. When surveillance was not possible (eg, in a traditional hospital service or at home), an appropriate medical device was used (Snootzz).
- (2) Nasopharyngeal airway tube: The NT was used to relieve obstruction of the upper airways since 2001. The purpose of this NT is to bypass the base of the

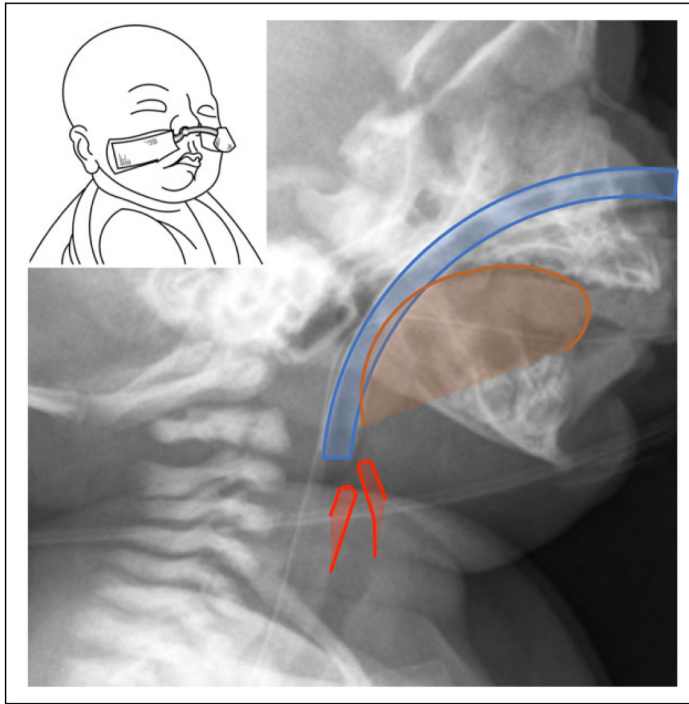


Figure 1. Schematic representation of the effect of the nasopharyngeal tube in a profile newborn radiography and of a newborn with a nasopharyngeal tube.

tongue, the site of obstruction of the upper airways in most cases in PRS. This tool is a major element of our therapeutic sequence and is systematically tested in the treatment of acute respiratory distress of the newborn with suspicion of PRS, repeatedly avoiding tracheal intubation.

NT's indications were as follows:

- Obstructive symptoms (snoring/noisy breathing, labored breathing, retractions, hyperextended neck in prone position) during awake clinical examination;
- Pathological respiratory polygraphy (RP) in prone or lateral position (see below for details).

The Portex Uncuffed Ivory PVC tube (Smiths Medical) with a diameter of 3.5 mm was used. The tube was inserted into the largest nostril hole until the resistance of the passage from the base of the tongue was felt by the surgeon or neonatologist. The average length of the tube is approximately 7 cm (see Figure 1). The rest of the outer tube was longitudinally divided to fix the strips on the baby's cheeks with Duoderm (ConvaTec).

The correct positioning of the tube was confirmed by a lateral radiological examination. The tip of the tube should be placed immediately under the base of the tongue without touching the upper part of the epiglottis. The relief of upper airway obstruction has been clinically evidenced by the disappearance of obstructive noises and breathing difficulties. Since 2010, RP was performed with the NT in place to control the release of obstructive breathing during sleep and obstructive

apnea-hypopnea index (OAH). Respiratory polygraphy is repeated at least 48 hours after removing the NT.

- (3) Surgical treatment: Before 2001, surgery was performed in the absence of relief of respiratory distress with NT. No mandibular distraction or tongue lip adhesion was used in this indication in our center.

A simplified version of the system of classifying babies with PRS based on the severity of their symptoms and signs and treatment required proposed by Cole et al. (2008) was used:

- Group 1: children requiring no specific respiratory treatment.
- Group 2: children requiring prone or lateral positioning.
- Group 3: children requiring NT and/or surgical treatment.

Feeding Support

Suction and swallowing may be impaired in PRS secondary to the obstructive problem or by immaturity of the function, so the support of enteral feeding appeared as an associated indicator of the severity. Support for enteral feeding has been indicated when the oral diet alone was unable to achieve normal weight gain. The duration of the use of the nasogastric tube and/or the gastrostomy, the “age of exclusive oral feeding,” the day when exclusive oral alimentation was sufficient to ensure normal weight gain (the day when nasogastric tube or gastrostomy was removed successfully), and the first day of life (noted day 0) for children who did not required feeding support at all were collected.

Respiratory Polygraphy

Respiratory polygraphy, instead of polysomnography, in PRS was practiced since 2010 in case of suspected obstructed breathing. Indeed, RP is considered by the clinical consensus report as an adequate sleep assessment in infants with PRS (Breugem et al., 2016). However, RP does not include electroencephalogram monitoring and thus the presence of obstructive hypopneas will be underestimated by this technique. Respiratory polygraphy includes a measurement of oxygen saturation, transcutaneous carbon dioxide, cardiac and respiratory frequency, nasal flow, and thoracic movements. The scoring of respiratory events follows the recommendations of the American Academy of Sleep Medicine (Berry et al., 2012). In case of clinical urgent upper airway obstruction, the respiratory treatment (prone or lateral positioning of NT) was started without waiting for the RP. In these cases, RP was performed during the treatment (eg, with NT in place) or at the end of the treatment, at least 48 hours after removal of NT, to certify that the obstruction of the upper airway was relieved.

The RP was considered abnormal if the OAH is >1 as recommended by van Lieshout et al. (2014) and Anderson et al. (2011).

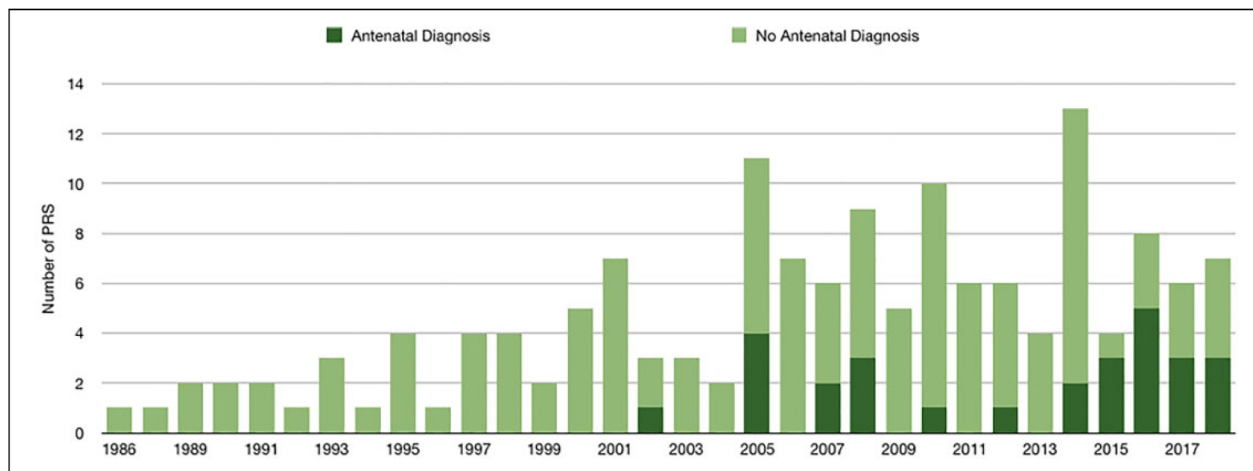


Figure 2. Evolution of the prenatal diagnosis of Pierre Robin sequence from 1986 to 2018.

Age at Surgery for Cleft Palate Repair

In PRS, surgery for cleft palate repair is only possible in children with satisfactory weight gain and after resolution of the upper airway obstruction. In our center, cleft palate repair is planned routinely around 6 to 8 months up to 12 months of age (we are not talking of isolated cleft palate in this article). We recorded the age at surgery, considering that an early intervention would demonstrate a success in treatment of the upper airway obstruction.

Statistical Analysis

The comparison of potential confounding factor (term of birth, birth weight, clinical presentation, and age of exclusive oral feeding) between the 3 groups was evaluated. Fisher test was used to compare the terms of birth and clinical presentations. Analysis of variance was used for the comparison of birth weights and that of Kruskal-Wallis for the ages of exclusive oral feeding.

Results

Clinical Data

The data of 150 PRS children (75 girls; 75 boys) were included. The prenatal diagnosis increased since 2002 (Figure 2). The birth was at term for 66% ($n = 99$) of the infants. The average birth weight was 3016 ± 623 g, the average birth size was 48 ± 4 cm, and the average head circumference at birth was 33 ± 2 cm. The Apgar score was normal for 96% ($n = 144$) of them. For the 6 infants confronted with initial respiratory distress, 2 required an urgent endotracheal intubation (born and initially managed in a peripheral center), 3 benefited from an immediate NT insertion, and 1 showed quick clinical improvement after lateral positioning. No mortality was observed during the first 6 months of life in either of these infants. Considering the age at the cleft palate repair, two-third of the cohort

Table 1. Description of Children With Syndromic Pierre Robin sequence.

Syndrome	n	Percentage of syndromic PRS	Percentage of total PRS
Stickler	8	36.4	5.3
22q11	2	9.1	1.3
Fragile X	2	9.1	1.3
Van der Woude	1	4.5	0.7
Achondrodysplasia	1	4.5	0.7
Kniest	1	4.5	0.7
Ehlers Danlos	1	4.5	0.7
Cerebro-costomandibular	1	4.5	0.7
Other chromosomal anomaly ^a	5	22.7	3.3
Total	22	100.0	14.7

Abbreviation: PRS, Pierre Robin sequence.

^a49 XXXXY, 46 XY 13p+, XXX, Xq28 duplication, partial monosomy Xp.

($n = 99$) underwent surgery before 9 months, and 89% ($n = 134$) before 12 months. The median age at surgery was 7.2 ± 3.4 months.

Regarding severity, 46% ($n = 63$) were classified in group 1, 36% ($n = 50$) in group 2, and 18% ($n = 25$) in group 3. In the latter group, mouth floor release described by Delorme et al. (1989) was performed for 7 children, cephalic traction for 3, and the use of an NT for 22 since 2001. No child has required a tracheostomy. Eighty (53%) children benefited from the use of a nasogastric tube. Thirteen (8%) required a gastrostomy. The median duration of food support was 3.5 days (0, 26.5; first and third quartiles).

After exploration and genetic counseling, 65% ($n = 97$) presented iPRS; 15% ($n = 22$) were syndromic. Stickler syndrome was the most common syndrome ($n = 8$; for more details, see Table 1). Twenty-one percent ($n = 31$) had PRS associated with other birth defects. Among these, 4 cases of PRS associated with a congenital clasped thumb were found.

Table 2. Confounding Factors of Initial Invasive Respiratory Treatment.

		Total		Group 1		Group 2		Group 3		P
		n	%	n	%	n	%	n	%	
Clinical presentation	Isolated	97	64.67	52	73.24	30	60.0	15	51.72	.07
	Associated	31	20.7	9	12.7	15	30.0	7	24.14	
	Syndromic	22	14.63	10	14.8	5	10.0	7	24.14	
	Total	150	100.0	71	100.0	50	100.0	29	100.0	
Term birth	At term	100	71.4	47	72.3	35	71.4	18	69.2	.91
Birth weight (g)	Mean (\pm SD)	3015.9 (\pm 622.56)		2917.3 (\pm 653.34)		3055.1 (\pm 600.63)		3159.6 (\pm 578.34)		.22
	Total (n)	131		57		48		26		
Day of exclusive oral feeding	Median (Q25, Q75)	3.0 (0.0, 27.0)		1.0 (0.0, 4.0)		10.5 (1.0, 28.0)		51.5 (30.0, 90.0)		<.01
	Total (n)	119		59		42		18		

Abbreviations: SD: standard deviation, Q25, first quartile, Q75, third quartile.

Confounding Factors of Initial Invasive Respiratory Treatment

Term birth, birth weight, or associated birth defects were not significantly different between the 3 groups (see Table 2).

The age of exclusive oral feeding was significantly different: 1 day (quartiles: 0-4) for group 1; 11 days (quartiles: 1-28) for group 2; 52 days (quartiles: 30-90) for group 3 ($P < .01$).

Efficiency of the NT

Respiratory polygraphy wasn't performed systematically before, during, or after NT utilization. Five children never get RP, and obstructive evaluation was made on clinical symptoms only (disappearing of obstructive noises with the NT, disappearing of the contraction of accessory respiratory muscles, observation of weight gain).

The NT was kept during 3.9 ± 2.4 months (minimum 1 week, maximum 10 months). Respiratory polygraphy was performed with NT to evaluate the respiratory obstruction relief in 10 children and after removing of NT in 4 children. The NT was not associated with any complication, in particular no ulceration of the nostril or of the nasal septum.

The length of hospital stay was not statistically different before the use of the NT in clinical routine and after 40 days ($n = 34$) and 35 days ($n = 116$), respectively, $P = .12$.

For 3 children, NT wasn't sufficient to relieve the upper airway obstruction. Two of them underwent cephalic traction in prone position followed by floor mouth release, respectively, at day 8 and day 15. One other infant boy had a severe gastro-esophageal reflux disease. Respiratory polygraphy at day 15 found obstructive events (OAHl = 5.5). Pediatric otorhinolaryngologist didn't found any laryngeal or tracheal anomaly after endoscopy. Nissen fundoplication was performed and finally the respiratory obstruction was relieved. Genetic examinations diagnosed later chromosome Xq28 duplications encompassing methyl-CpG-binding protein 2 gene syndrome, which is known to associate severe neuro-developmental disorder with hypotonia and hypersalivation.

The modifications of the results of the RP with NT for the 14 other children is presented in Table 3.

Discussion

This study describes the evolution of the treatment of the obstructive respiratory symptom in a large cohort of children with PRS in a single tertiary center. In the first period, severe obstruction was treated by prone positioning, oral surgery with or without NT. In the second period, the NT alone was the first line of the treatment in severe obstruction (group 3). In our experience and through the use of the NT, the second line of the severe obstruction, tracheostomy, was not used. The severe obstruction was significantly correlated with initial swallowing disorder requiring nasogastric tube and/or gastrostomy, esophageal reflux, and a later age of exclusive oral alimentation.

This cohort presents similar characteristics with previously reported series. The ratio of iPRS and nonisolated PRS varies between the study as a function of the definition of PRS used by the authors. We have a relatively high percentage of iPRS (65%) probably due to the fact that we include only PRS with a cleft palate. This higher rate of iPRS is reported in other study using the same definition (Morice et al., 2018). Moreover, sPRS was defined after a physical examination, malformative evaluation (eg, transfontanelle ultrasonography, echocardiogram, etc) and "fast results" basic genetic evaluations, at the age of 6 months. This early evaluation may lead to underestimate the rate of nonisolated PRS. In our cohort, sPRS or aPRS was not found to be associated with more severe respiratory treatment. However, Vipulanathan et al. (2014) reported that the presence of a cleft palate was associated with more respiratory failure.

The definition of PRS is variable in the literature (van Nunen et al., 2018) so in 2014, an international consensus meeting published a clinical consensus report based on a synthesis of the available literature and expert opinions (Breugem et al., 2016). They defined PRS as the association of micrognathia, glossoptosis, and upper airway obstruction. They provided clinical and nonclinical validated methods for the assessment of the severity of these features. This will lead to

Table 3. Consequences of the Use of Nasopharyngeal Tube on the Respiratory Polygraphy in 14 Children.

Patient number	Age when performing RP (days)	Presence of NT	Results of RP (normal or abnormal)	OAHl (when available)	Comments
1.	11	X	Normal	0.5	2 days after removing of NT
	120	–	Normal	0.5	
2.	7	X	Normal	0	
3.	8	–	Normal	0.5	
4.	10	–	Abnormal	10	No awake obstructive clinical symptoms
	60	–	Normal	0.5	2 days after removing of NT
5.	21	–	Abnormal	10	No awake obstructive clinical symptoms
	94	–	Normal	0.2	5 days after removing of NT
6.	10	X	Normal	1	No OSAHS but polypnea and desaturation due to cardiac malformation
7.	12	–	Abnormal	5	No awake obstructive clinical symptoms
	43	X	Normal	0.3	
8.	20	X	Normal	0.5	
9.	15	X	Normal	0.7	
10.	20	X	Abnormal	2	24 hours after removing of NT leading to repositioning the NT
	35	–	Abnormal	4.2	
11.	48	X	Normal	0.1	No awake obstructive clinical symptoms We tolerated 1,6 OAHl considering the dramatic shift obtained with NT
	5	–	Abnormal	15	
	9	X	Abnormal	1.6	
12.	11	–	Abnormal	–	Obstructive and central sleep apnea
	24	X	Normal	1	Normal RP after caffeine treatment and NT
13.	24	X	Normal	0.8	48 h after removing of NT
	60	–	Normal	0	
14.	14	X	Abnormal	2.6	Radiological examination found that NT was not well positioned
	26	X	Normal	0.6	NT well positioned

Abbreviations: NT, nasopharyngeal airway tube; OAHl, obstructive apnea-hypopnea index; OSAHS, obstructive sleep apnea-hypopnea syndrome; RP, respiratory polygraphy.

more homogeneous clinical groups of children having PRS with comparable and consistent treatment options (Côté et al., 2015). In this retrospective study, the diagnosis criterion defined in Utrecht was not used. The PRS was defined by the morphological triad (micrognathia, glossoptosis, cleft palate) and 46% of the children did not present symptoms of upper airway obstruction at least during their first 6 months of life. Perhaps, this is explained by the fact that this large cohort is extracted from a surgical database over a 30-year period (Pierre Robin, 1934).

The severity of the PRS differs also between studies. We chose to use a clinical simplified system of classification (adapted from Cole et al. [2008], dividing our cohort in 3 groups of severity. However, the PRS recognized as grade 1 or 2 at birth can worse in the first 1 or 2 weeks from birth becoming a grade 2 or 3. In these cases, the decision to insert the NT is made after 2 weeks postbirth. In most of these cases, there is an associated laryngomalacia that can worse the respiratory distress. The evaluation by a pediatric otolaryngologist is thus critical.

The most recent studies seem to show that nonsurgical management is preferred in PRS (Morice et al., 2018; Gómez et al., 2018b). This study confirms these data. Mandibular distraction

or tongue-lip adhesion was not performed in this cohort. However, the use of the NT was the first line to relieve the respiratory obstruction. The nurses are used to the management of children with NT and are able to teach the parents too. The use of NT is described already in the classification of the severity symptoms of PRS made by Benjamin and Walker in 1991. It has been recently showed that it allows to reduce the need for surgery in PRS (Gangopadhyay et al., 2012). Poets et al. (2017) in a prospective longitudinal cohort study of the efficiency of the nasopharyngeal airway don't find any case requiring switching to a surgical procedure. The NT is also promising because it allows sucking and swallowing function during the treatment. With good parental acceptance and education to the management of the NT (how to insert, remove, and clean it), it allows also to home management (Anderson et al., 2007). A RP was performed systematically, before returning home with the NT, to assess the total delivery of upper airway obstruction, even during sleep. Before going home, all the equipment was provided. The babies were monitored by electrocardiogram and pulse oximeter, 24 hours a day. A specialized nurse came to the house every day. Probe position, weight, occurrence of monitoring events, and comfort score were checked daily. Emergency medical assistance was available day and night if

needed, due to medicalized pediatric ambulance service in the region of the competence center.

A negative point is that in case of several levels of airway obstruction, the NT will not be sufficient. However, tongue-lip adhesion and mandibular distraction neither (Andrews et al., 2013). In these cases, only continuous positive airway pressure, tracheal tube, or tracheostomy would be an option (Essouri et al., 2005). If necessary, the second line of treatment after NT would be the tracheostomy. Another limitation of the use of nasopharyngeal intubation could be the difficulty to find available tubes designed directly for this purpose. Indeed, altering the Portex endotracheal tube could be a barrier in centers that have policies in place that prohibit such alterations of medical devices. Finally, *a contrario* of mandibular distraction which improves the upper airway patency but also creates a more typical appearance, with NT the appearance of the face is not immediately changed. This information has to be discussed with the parents, and information that the improvement in the small jaw with the NT approach in infancy depends on time and natural growth should be given.

German teams recently report great results of relieving the upper airway obstruction by the use of a more technical nasopharyngeal airway tool: the PEBP (Vatlach et al., 2014; Poets et al., 2017). The PEBP improves upper airway obstruction as well as weight gain of children with PRS (Buchenau et al., 2017; Poets et al., 2017). The used of NT go in the same way: treatment of the obstruction first to improve the alimentation and weight gain. Recently, a new study shows that this non-invasive treatment could promote mandibular growth (Wiechers et al., 2019). The PEBP presents the disadvantage that the teams have to be trained in its management, whereas the NT may be easily available in every maternity service. It cannot be used in emergency because of the time required for creating it by an experienced prosthetist and an orthodontist. The PEBP appears, however, to be a promising noninvasive tool for the treatment of PRS and maybe surpassing the NT. Prospective studies are still required to prove it.

The limits of this study are its retrospective nature that makes the design susceptible to significant bias. In most of the cases, especially before 2010, we used only clinical evaluation of the upper airway obstruction. Because RP was not systematically performed, we are not able, in most cases, to do a quantitative evaluation of our intervention. However, the good clinical evolution of these children attests that our algorithm works well. One example of this good evolution is the age of surgery for cleft palate repair. For patients of group 1: a normal weight gain and no obstructive symptoms (snoring/noisy breathing, labored breathing, retractions, hyperextended neck in prone position) during awake clinical examination were required before surgery. For patients of group 2 and 3: the same criteria than group 1 and a normal sleep RP was required before planning surgery. According to this, 89% of the patients of the cohort underwent surgery before 12 months of age, attesting a satisfactory, even not perfect, evolution.

Through this study, we described our algorithm of the initial respiratory treatment of children with PRS. First of all, as

mentioned in the clinical consensus report, we ensure the breathing, with conservative treatment as prone or lateral positioning, or NT. The normal breathing (normal oxygenation) is the main key to gain weight and for brain development of the children. In case of normal clinical examination without apparent upper airway obstruction during the time of awakening, we recommend to perform sleep RP. In clinical practice, we do recommend thus systematic RP assessment for all patients with PRS. After ensuring the breathing and a short period of suspension of oral feeding, we start slowly to feed the baby again. During all the time where NT is in place, sucking is encouraged and oral alimentation is possible.

Conclusion

Over time, NT became our first-line major treatment to relieve the upper airway obstruction, without surgery required for most of the children. In parallel, the age of exclusive oral feeding seems to be a good factor to predict the severity of the evolution.

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