



# Pierre Robin sequence: treatment with nasopharyngeal tube

Pierre Robinova sekvenca: zdravljenje z nosno-žrelnim tubusom

Mojca Železnik,<sup>1</sup> Andreja Eberlinc,<sup>2</sup> Daša Gluvajčić,<sup>3</sup> Uroš Krivec,<sup>4</sup> Jana Lozar Krivec<sup>1</sup>

## Abstract

Pierre Robin sequence is described as a triad of micro- and/or retrognathia, glossoptosis, and upper airway obstruction, in 90% the triad is associated with cleft palate. In children with the Pierre Robin sequence, the main functional problems are upper respiratory obstruction and feeding problems, which can be manifested by a variety of respiratory problems, and poor weight gain. Most patients need conservative treatment, rarely surgical treatment; only the most severe cases will need a tracheotomy. In Slovenia, at the Clinical Department of Neonatology, Division of Paediatrics – University Children's Hospital we introduced a new method for the treatment of newborns with the Pierre Robin sequence, treatment with the nasopharyngeal tube. The nasopharyngeal tube overcomes the obstruction of the upper respiratory tract at the level of the root of the tongue and can be placed for a longer period of time, even in the home environment; we can alternately replace the tube from one to another nostril. The treatment of children with the Pierre Robin sequence is multidisciplinary and involves a neonatologist, otorhinolaryngologist, maxillofacial surgeon and a pulmonologist, while a gastroenterologist, clinical dietitian, and geneticist are included if needed. The article presents the clinical picture and the management of children with Pierre Robin sequence, and the clinical pathway for the evaluation and treatment of the neonate with Pierre Robin sequence, which was introduced at the Department of Neonatology, Division of Paediatrics – University Children's Hospital, University Medical Centre Ljubljana.

## Izvleček

Pierre Robinova sekvenca je triada, ki jo sestavljajo mikro- in/ali retrognatija, glosoptoza in zapora zgornjih dihal, ki se ji v 90 % pridruži palatoshiza. Pri otrocih s Pierre Robinovo sekvenco sta glavni funkcionalni težavi obstrukcija zgornjih dihal in oteženo hranjenje, ki se kaže z raznolikimi dihalnimi težavami in slabim pridobivanjem telesne mase. Pri večini bolnikov

<sup>1</sup> Department of Neonatology, University Children's Hospital Ljubljana, University Medical Centre Ljubljana, Ljubljana, Slovenia

<sup>2</sup> Department of Maxillofacial and Oral Surgery, University Medical Centre Ljubljana, Ljubljana, Slovenia

<sup>3</sup> Department of Otorhinolaryngology and Head & Neck Surgery, University Medical Centre Ljubljana, Ljubljana, Slovenia

<sup>4</sup> Department of Paediatric Pulmology, University Children's Hospital Ljubljana, University Medical Centre Ljubljana, Ljubljana, Slovenia

**Correspondence / Korespondenca:** Jana Lozar Krivec, e: [jana.lozar@gmail.com](mailto:jana.lozar@gmail.com)

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**Ključne besede:** Pierre Robinova sekvenca; novorojenček; zapora dihal; nosno-žrelni tubus

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zadoščajo konzervativni ukrepi, v redkih primerih pa je potrebno kirurško zdravljenje, v najtežjih primerih pa traheotomija. Klinični oddelek za neonatologijo Pediatrične klinike UKC Ljubljana je za zdravljenje novorojenčkov s Pierre Robinovo sekvenco uvedel v slovenski prostor novo metodo, tj. vstavitve nosno-žrelnega tubusa. Ta premosti zaporo zgornjih dihal v višini korena jezika in je ob izmenični zamenjavi strani vstavitve skozi nosnici lahko nameščen daljše obdobje tudi v domačem okolju. Obravnava otrok s Pierre Robinovo sekvenco je multidisciplinarna. Pri obravnavi sodelujejo neonatolog, otorinolaringolog, maksilofacialni kirurg in pulmolog. Gastroenterolog, dietetik in genetik pa so vključeni po potrebi. Članek predstavi klinično sliko otrok s Pierre Robinovo sekvenco, metode zdravljenja in uradno klinično pot obravnave v Sloveniji.

## 1 Introduction

Pierre Robin Sequence (PRS) is a condition characterized by a sequence of lower and upper jaw abnormalities and thus respiratory and gastrointestinal problems (1). In 1923, the French dentist Pierre Robin described a triad consisting of micro- or/and retrognathia, posterior displacement of the tongue into the oropharynx and hypopharynx (glossoptosis), and thus the obstruction of the upper respiratory tract. In 90% of cases, the triad is associated with a cleft palate (palatoschisis) (2-4) (Figure 1).

The estimated incidence ranges from one in 8,000 and 20,000 births, depending on the PRS definition (2,3,5,6). Airway obstruction is a major predictor of morbidity and mortality. The mortality rate of all children with PRS ranges from 1.7% to 11.3%, and in patients with associated other abnormalities it is up to 26% (3-5).

## 2 Aetiology

The primary pathogenetic event leading to PRS is unknown. Researchers believe that hypoplasia of the lower jaw, which occurs before the 9th week of gestation, causes the tongue to move up and back, which mechanically prevents the fusion of the palatal sutures between the 8th and 10th week of gestation (3,4,7,8).

PRS can manifest either as isolated abnormality in the development of the oropharyngeal area or in 45-80% of cases there are other anomalies associated, PRS being a part of the syndrome. Therefore, it is advisable that a clinical geneticist is involved in the management of a neonate with PRS, who, if necessary, performs further genetic tests (3-5,9). Most often, PRS is associated with Stickler's syndrome, which is a connective tissue disease and also the main reason for retinal detachment in children (7).

## 3 Pathophysiology

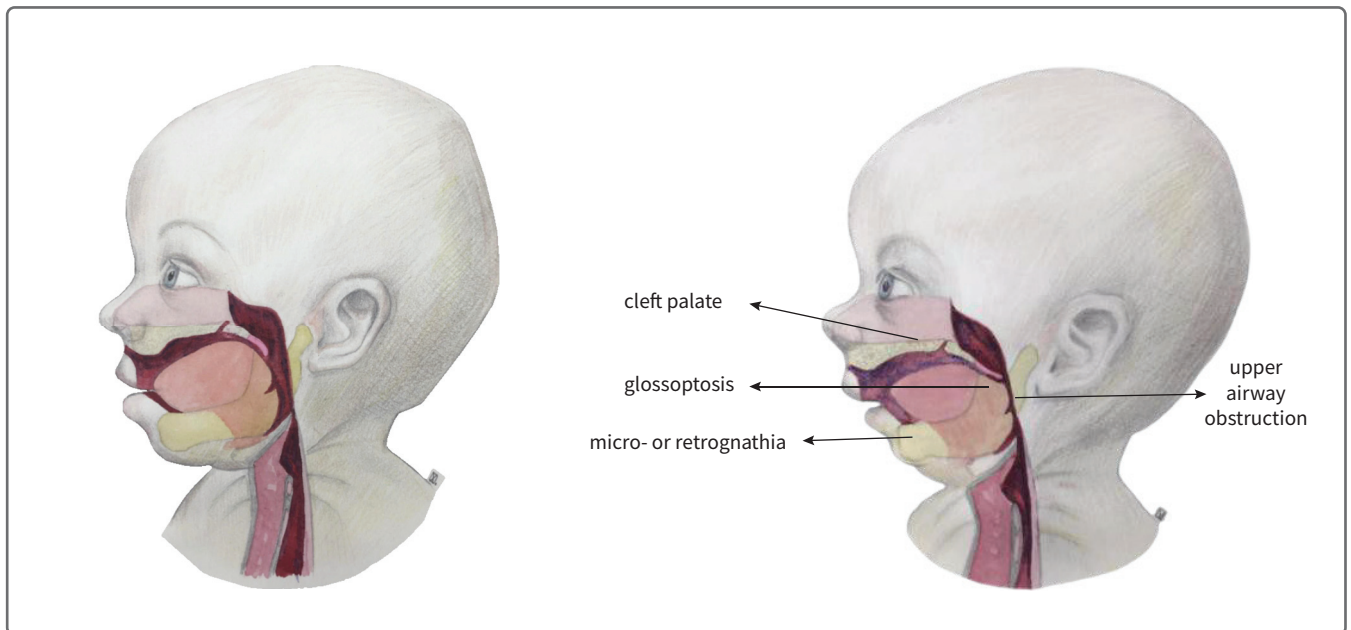
In a child with PRS, the main functional problems are dynamic upper airway obstruction and feeding disorder, which cause breathing disorders and poor weight gain (2). The degree of upper airway obstruction, which

occurs primarily due to micro- or retrognathia and airway obstruction with the root of the tongue, is not constant (5,7) (Figure 1). The tongue compressed inside the small lower jaw, the tongue prolapses into cleft palate, the lack of control over the muscles of the tongue, and the pressure of the tongue on the oropharynx and hypopharynx further contribute to upper airway obstruction. Obstruction usually decreases over time, but it is not clear whether the cause of the improvement is faster growth of the lower jaw after birth or a decrease in glossoptosis with growth and neurological development (3,8). Additional factors that may contribute to airway obstruction in children with PRS are described, such as: hypotonia, central sleep apnoea, laryngomalacia, tracheomalacia, and subglottic stenosis (3).

The causes of feeding problems are many: from micrognathia, glossoptosis, concomitant cleft palate, impaired mobility of the tongue, to oesophagus motility disorder, which causes gastroesophageal reflux (5). Increased work of breathing and tachypnoea negatively affect the coordination of sucking, swallowing and breathing. Feeding problems increase the possibility of aspiration and pulmonary complications (5,10). Gastroesophageal reflux affects the balance between respiration and feeding, as it causes inflammation and swelling of the airways, increases secretions and thus influences the mechanics of swallowing (3). Associated heart or other anomalies may further contribute to feeding problems and thus to poor weight gain (5).

## 4 Clinical picture

Depending on the degree of upper airway obstruction, children with PRS may have a number of signs of respiratory distress that appear immediately after birth or in the first weeks of life (2,3). Before birth, the anomaly is rarely visible. Micrognathia can be seen by ultrasound or magnetic resonance imaging, and in the case of glossoptosis and swallowing disorders in the foetus, ultrasound can detect polyhydramnios (3,11,12).



**Figure 1:** Schematic presentation of a neonate with normal anatomy of the upper respiratory tract (left) and with Pierre Robin sequence (right).

After birth, upper airway obstruction is often at the forefront, manifested by grunting, breathing difficulty, and obstructive sleep apnoea, as well as feeding difficulties. Glossoptosis and airway obstruction can be of varying degrees. In severe obstruction, patients may have inspiratory stridor or biphasic stridor, and occasionally even cyanosis (8,9), whereas in mild glossoptosis the airway may be clear in the waking state and during suckling, but during sleep, obstructive sleep apnoea may occur (13). Early recognition and intervention are key to prevent the consequences of breathing disorders such as: poor weight gain, food aspiration, respiratory failure, persistent hypoxaemia, hypercapnia, gastroesophageal reflux, increased pulmonary vascular resistance, and thus the development of cor pulmonale condition, neurological impairment and also death (14).

Cleft palate is present in 90% of children with PRS, therefore a large proportion of children have problems with establishing a normal feeding pattern due to suboptimal anatomical conditions because of the communication between the oral and nasal cavities, which prevents the creation of intra-oral vacuum, necessary for successful suckling (15-17). A recently published Slovenian study of children with orofacial clefts showed that almost three quarters of children had problems establishing feeding immediately after birth, and more than a seventh of them the problems persisted even after surgical treatment (15).

The cleft palate also affects speech development and speech disorders later on (articulation disorders and hypernasality), more than half of children with palatoschisis

also have recurrent otitis media or otitis media with effusion with conductive hearing loss (18). Interventions such as a tracheotomy in a child with PRS can have an additional effect on speech development (19).

## 5 Diagnostics

In 2020, the consensus of paediatricians and otorhinolaryngologists on the recommended treatment of children with PRS was published (20). In order to select the optimal treatment, in addition to the clinical assessment of breathing effort during wakefulness, sleep and feeding, it is necessary to evaluate the level of respiratory disturbances and determine the location of airway obstruction. The association between the degree of micrognathia and the degree of glossoptosis has not been demonstrated, which means that the assessment of micrognathia cannot predict the degree of airway obstruction (21). Before starting treatment, endoscopic airway examination is advised to assess glossoptosis and location of airway obstruction (6,11). With flexible nasolaryngoscopy, the upper respiratory tract and vocal cord mobility can be assessed, and the presence of laryngomalacia can be demonstrated. The disadvantage of this examination is that the child is usually restless and crying during the examination, which can make the assessment of the level and the degree of airway obstruction due to glossoptosis, which is most pronounced when the child is asleep, impossible (22). Flexible nasolaryngoscopy and/or bronchoscopy in a sedated child provide a better assessment of the location of airway

obstruction, and allows additional assessment of the entire airway, thus identifying any attendant abnormalities such as subglottic stenosis, tracheomalacia, bronchomalacia and stenosis of trachea (3,11).

Using cardiorespiratory polygraphy during sleep in the first months after birth allows an objective evaluation of breathing disturbances (2,3,23). Obstructive sleep apnoea is extremely common in patients with PRS. A retrospective study by Hicks et al., which included 31 neonates with PRS, found moderate to severe obstructive sleep apnoea in 64% of neonates (6). Cardiorespiratory polygraphy during sleep also allows to evaluate the effectiveness of the measures taken in the patient and to assess the improvement or worsening of the level of obstruction (6,8). If left untreated, the obstruction usually progresses in the first 4–6 weeks after birth (3).

When assessing the feeding of children with PRS, it is important to distinguish between feeding problems due to upper airway obstruction and problems resulting from altered anatomy in the presence of cleft palate, swallowing disorders, aspirations, and gastroesophageal reflux. Additionally, feeding and growth problems may occur in connection with the underlying syndrome or other associated abnormalities (3). Clinical diagnostics of gastroesophageal reflux can be difficult. A 24-hour pH impedance testing is most commonly used to quantify reflux (3,24,25). Inadequate weight gain can also result from increased calorific needs due to increased work of breathing. A clinical dietitian and a speech therapist must be included in the management, while a personal paediatrician is responsible for long-term monitoring of growth and development. A paediatric gastroenterologist is consulted in cases of severe gastroesophageal reflux disease and oesophageal motor dysfunction (3,11).

## 6 Treatment

There are many methods used to treat neonates and children with PRS. Although PRS is a well-known clinical entity, current classification systems do not consider the diversity of the clinical picture of neonates with PRS, that could allow the selection of the optimal treatment for a particular clinical case (3,4,26).

The protection of the airway is crucial in the treatment of children with PRS, since it enables easier breathing and feeding, and lowers the risk of food aspiration during oral feeding (5). A multidisciplinary approach is advised, which facilitate faster and more efficient identification of problems, prevents unnecessary interventions and enables optimal care for children with PRS (3).

In more than half of the children, conservative,

non-surgical interventions are sufficient (3). The first-choice measure, which is successful in more than half of patients with mild to moderate airway obstruction, is the prone or side position, which allows the lower jaw and tongue to fall forward, moving the tongue away from the back of the pharynx (4).

If this measure is not successful, we decide to insert a nasopharyngeal tube (NPT) or use respiratory support with continuous positive airway pressure (CPAP) or non-invasive ventilation (NIV) (3,21). According to a study by Abel et al., the effectiveness of NPT in children with PRS with moderate to severe obstruction is 80%, and the use of NPT also reduces the number of required tracheotomies (8). In a study by Leboulanger et al., the use of NIV reduced the proportion of time of SpO<sub>2</sub> below 90% from an average of 14% to 1%, and also reduced the value of transcutaneously measured partial pressure of carbon dioxide (P<sub>tc</sub>CO<sub>2</sub>) from 7.6 to 4.1 kPa (9). The disadvantages of NIV are: the difficulty in optimal mask selection due to the changed facial anatomy, the difficulty of establishing oral feeding, and ventilation intolerance (9,23). When technical limitations or inability to accept ventilation support enable the use of CPAP or NIV, at least some degree of respiratory support can be achieved with high-flow therapy (HFT) (27).

Some centres use custom-made orthodontic plates (e.g. a pre-epiglottic baton plate). The plate reduces upper airway obstruction by resolving glossoptosis. It is used mainly in children with a milder form of PRS (2).

Among surgical methods of treatment, the most common are surgical attachment of the base of the tongue to the lower lip (glossopexy) and mandibular distraction osteogenesis. The first is indicated when the patient has severe glossoptosis, and endoscopy has ruled out subglottic stenosis or other respiratory abnormalities, and it consists of two procedures (5,11). The secondary procedure, which allows the base of the tongue to be released, is usually performed between 12 and 18 months of age (11). According to studies, the effectiveness of the method for resolving the airway obstruction is between 71% and 89% (4,11). Recurrence of glossoptosis may occur after adhesion release; other complications of the procedure include wound dehiscence, tongue swelling after surgery, and scarring (2,4,5).

Mandibular distraction osteogenesis consists of bilateral vertical osteotomy of the hypoplastic mandibula followed by an external or internal distractor placement (2,4). The external distractor is easier to place and remove. Possible disadvantages of its use are the movement of the distractor due to the external forces and scarring at the site of its placement. The internal distractor is usually

better tolerated by children, but secondary surgery under general anaesthesia is required for its removal. Osteotomy is followed by an activation phase, when the bone is stretched and it grows at a rate of 0.5–2 mm per day. The final stage is consolidation, when the newly formed bone is ossified (3). Until the lower jaw sufficiently grows, the airway must be secured with a NPT or an endotracheal tube, or the patient should breathe through a tracheostomy (2-4). Mandibular distraction osteogenesis allows rapid resolution of obstruction, but, like any surgical treatment, has certain risks (2,4). Compared with glossopexy, it allows faster establishment of oral feeding (5), while other authors observe poor weight gain as a result of dysphagia, which is not corrected after distraction (2). A possible complication of treatment may be damage to the mandibular and inferior alveolar nerve, and damage to the molar design, which occurs in half of the cases (2,4,6). Mandibular distraction osteogenesis is probably most effective when micrognathia and glossoptosis are at the forefront. However, in the case of multilevel airway obstruction, distraction will not be effective and patient will require a tracheotomy despite elongation of the lower jaw (6).

Tracheotomy does not correct basic PRS abnormalities, but it is the only method that efficiently resolves airway obstruction. It is an invasive method and is used in children with PRS when: other methods of treatment are not successful, in very severe airway obstruction, in multilevel airway obstruction, and in patients with associated anomalies (2,3,23,28). In children with tracheotomy the hospitalization is longer. In 65% of cases there are complications associated with tracheotomy, which can also be life-threatening (19,28). The most common complications are: bleeding, pneumothorax, decannulation, tracheitis, tracheal stenosis, and, exceptionally, even sudden death (2,3).

90% of children with PRS have palatoschisis, which does not require specific treatment in the first months of life, except for an adapted bottle and an adapted feeding position (15). According to the current recommendations, an isolated cleft palate is treated by maxillofacial surgeons after the first year of life (29,30).

Regardless of the treatment method, 38–62% of neonates with PRS have such severe feeding difficulties that a gastric tube must be inserted, either orally or nasally (3). The use of gastric tube is recommended in all patients with feeding difficulties and it does not interfere with the decision for discharge into home care. It is usually needed for a transitional period (3). The gastric tube provides additional benefits by further opening the airway, as it allows the tongue to be displaced away from the posterior

pharyngeal wall, and by reducing air swallowing during feeding, which can cause an increase in gastric pressure and thus contribute to gastroesophageal reflux. Placing the baby in an upright position is recommended to prevent reflux. If this measure is not sufficient, other anti-reflux measures, including the introduction of a proton pump inhibitor, may be advised (3,10). Children with associated syndromic disease or other anomalies and neurological disorders may have chronic feeding problems that require gastrostomy tube insertion (3,11).

After discharge from the hospital, the child's paediatrician plays an important role in following a child with PRS by paying particular attention to breathing and feeding problems. He/she needs to monitor the child's growth and development. In children with cleft palate, special attention must be paid to hearing and speech development (2,3,8,22). In most cases, children with PRS have a good prognosis, and timely prevention of hypoxic events allows them normal cognitive development (3).

Despite the progress in management, analyses of the results of different ways of treating children with PRS are lacking, so the approach to a child with PRS depends on the experience and strategy of specific centre. However, the international recommendations of paediatricians and otorhinolaryngologists on the management of children with PRS from 2020 allow several management approaches (20).

## 7 Pierre Robin Sequence treatment with a nasopharyngeal tube

Until now, the Department of Neonatology has used conservative treatment methods in neonates with PRS and mild to moderate signs of airway obstruction, such as prone or side position and insertion of a gastric tube or CPAP therapy. In patients with severe airway obstruction, tracheotomy was used, while glossopexy and mandibular distraction osteogenesis were not used in agreement with maxillofacial surgeons due to a number of possible side effects. In 2020, based on positive experiences of foreign centres, we introduced NPT treatment as a possible method of maintaining a free airway.

Table 1 shows the number of children born with schisis, palatoschisis, and PRS between 2016 and 2020 in Slovenia. According to the five-year period, the incidence of PRS in Slovenia is 1.2 per 10,000 live births. Of the 12 children born with PRS, seven children needed a gastric tube, two children born in the last year were managed with NPT, and one child required a tracheotomy.

After preparing the patient, the NPT is inserted through the nose, without general anaesthesia, so that

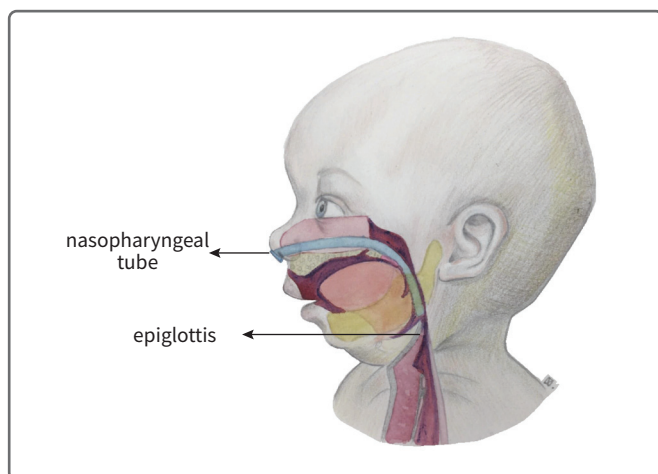
**Table 1:** Number of all children born with schisis, palatoschisis and Pierre Robin sequence between 2016 and 2020 in Slovenia.

Year of birth	No. of children born with schisis	No. of children born with palatoschisis	No. of children born with PRS
2016	27	10	3
2017	26	12	0
2018	33	12	3
2019	22	9	3
2020	25	11	3

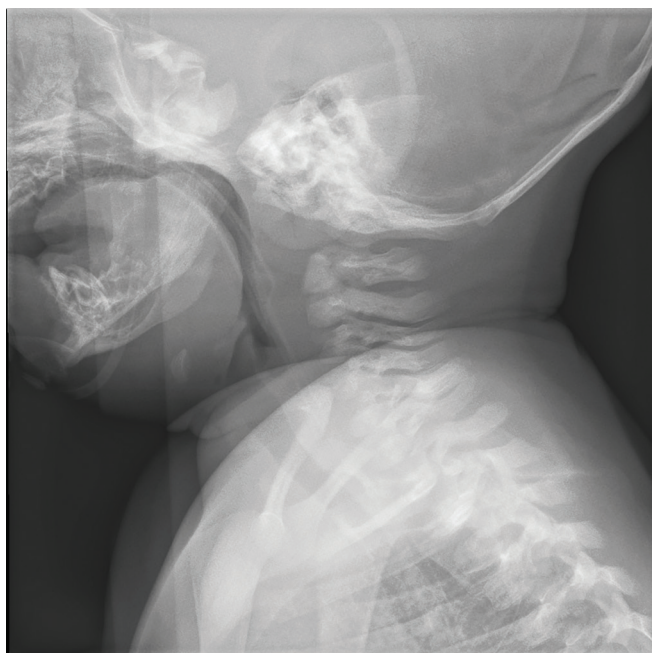
Legend: No. – number; PRS – Pierre Robin sequence.

the distal tip of the NPT is positioned behind the root of the tongue, but above the epiglottis (2,3,6). The intention of the insertion of NPT is to overcome the airway obstruction at the level of the oropharynx or root of the tongue and facilitate breathing and feeding. A properly inserted NPT allows the child to breathe through both, the tube and the free nostril, as the tongue is moved away from the posterior pharyngeal wall due to the inserted NPT (3) (Figure 2). Given the way in which obstruction is resolved, the use of NPT is probably the most effective method when the cause of airway obstruction is micro- or retrognathia and glossoptosis (2).

The depth of NPT insertion is determined by measuring the distance between the nostril and the ear tragus of the patient or by direct visualisation of the NPT tip with a flexible endoscope. The insertion depth can also be determined using the tables that determine the size



**Figure 2:** Schematic presentation of the inserted nasopharyngeal tube.



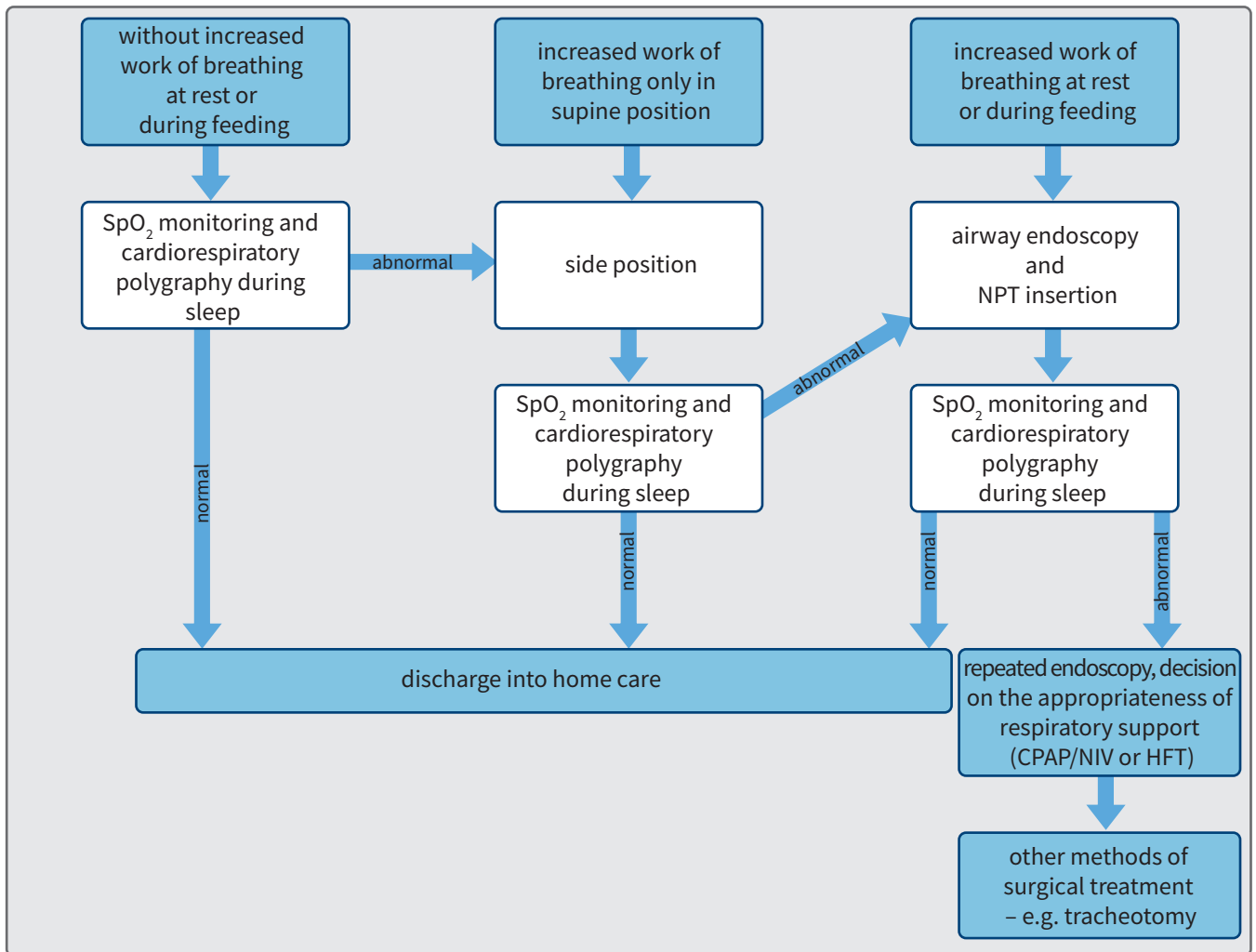
**Figure 3:** Lateral X-ray image of the head in the neutral position; the tip of the inserted nasopharyngeal tube is at the level of the epiglottis; the position of the nasopharyngeal tube should be corrected.

and depth of the insertion of the NPT according to body weight (11,14,31). After the first insertion of the NPT, the correct position can be confirmed by lateral neck X-ray in the neutral position of the head (8) (Figure 3).

The child has an NPT inserted 24 hours a day for weeks or months. Discharge from the hospital to the home environment is possible when parents learn to remove and replace, aspirate, and fix the NPT, as well as to handle a child with an inserted NPT (3). Masters et al. anticipate changing the NPT every 2–4 days at first and later on every 5–7 days (14). On average, children need NPT for 3–4 months or until the airway obstruction is relieved due to enlargement of the lower jaw which is the result of rapid growth after birth, stimulated mainly by suckling (3,6,11).

NPT treatment can have complications which are, nevertheless, rare. Complications include blockage of the NPT with secretions, aspiration of gastric contents due to incorrect positioning, inadvertent removal of the NPT, damage to the skin, cartilage and mucosa of the nose, and the formation of nasal stenosis (2,3).

At the Department of Neonatology, based on our own experience and professional reports, we have developed a protocol for the treatment of neonates with PRS with NPT (Figure 4). Indications for NPT insertion are: clinically perceived airway obstruction and signs of increased work of breathing at rest, significant sleep desaturations or obstructive episodes detected by the cardiorespiratory



**Figure 4:** Clinical pathway for treatment of neonate with Pierre Robin sequence.

Legend: NPT – nasopharyngeal tube; CPAP – continuous positive airway pressure; NIV – non-invasive ventilation; HFT – high-flow therapy.

polygraphy, feeding problems and poor weight gain, and increased work of breathing with hypercapnia despite the neonate being in the prone<sup>1</sup> or side position. Contraindications for the insertion of NPT are damage to the nose and throat, obstruction of the nasal cavity, septal deviation, blood clotting disorders, and additional airway obstruction below the epiglottis. Before the insertion of NPT, cardiorespiratory polygraphy while sleeping is performed. The size and approximate depth of the NPT insertion is determined using tables. The first insertion of NPT is performed in sedation during a flexible nasolaryngoscopy, which, in addition to assessing the upper respiratory tract, nasal patency, vocal cord mobility and location of airway obstruction, enables accurate insertion of the NPT just above the epiglottis, between the base of

<sup>1</sup>The prone sleeping position is a risk factor for sudden, unexpected infant death, so the patient should be monitored while in this position.

the tongue and the posterior pharyngeal wall. After insertion of the NPT, a lateral neck X-ray is performed in the neutral position of the head. If necessary, the depth of insertion of the NPT is corrected to ensure the tube insertion 1 cm above the epiglottis (Figure 3).

After insertion of the NPT, vital signs are continuously monitored, especially SpO<sub>2</sub>, and a cardiorespiratory polygraphy is performed during sleep. Patients are bottle-fed using special bottles and nipples for patients with schisis, and if necessary, a gastric tube is inserted. Evaluation of feeding or oromotor functions is performed according to NOMAS® (Neonatal Oral - Motor Assessment Scale) (32). Antireflux measures are taken in all patients, such as upright position and pre-thickened formula milk or feed thickener. If these measures are not sufficient, a proton pump inhibitor is introduced. After insertion of the NPT, the patient is still placed in a prone or side position. We recommend changing the NPT every 1-2 days



**Figure 5:** Photograph of a child with a nasopharyngeal tube inserted (the child's parents have agreed to publishing the article containing their child's photograph).

at first, then on every 3-5 days, alternately in one or the other nostril. If necessary, the size of the NPT is changed as the child grows.

During hospitalization, parents are taught to remove and replace the NPT, aspirate the tube and upper

respiratory tract, and feed the child by gastric tube if necessary. Parents are taught basic resuscitation procedures and are made aware of emergency measures in case of suffocation or food aspiration. Before discharge to the home environment, we inform the child's paediatrician and the visiting nursing service about the patient's condition and special medical needs and care. We introduce the child to the pulmonologist, as the infants continue their management at the Department for Pulmology of the University Children's Hospital, University Medical Centre Ljubljana (Figure 5).

## 8 Conclusion

PRS is a rare, life-threatening clinical condition. It requires timely identification and action. At the Department of Neonatology, a new method for the treatment of children with PRS by inserting NPT was introduced, which by resolving the obstruction at the level of the pharynx and base of the tongue allows maintaining a free airway in children with PRS.

## Conflict of interest

None declared.

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