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Care and conservative treatment methods for infants with Pierre Robin sequence

Metody pielęgnacji i leczenia zachowawczego niemowląt z sekwencją Pierre'a Robina

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Abstract

Introduction: Pierre Robin sequence is a congenital developmental disorder the aetiology of which is not fully understood. The prevalence of Pierre Robin sequence ranges from 1:8,500 to 1:20,000 live births, with mortality estimated between 3.6% and 21%. Anatomic malformations present in patients with Pierre Robin sequence cause airway obstruction and difficulties in oral feeding. **Aim:** The aim of this study was to summarise the current state of knowledge on conservative treatment methods and appropriate care for patients with Pierre Robin sequence. **Materials and methods:** We conducted a literature review on non-invasive treatment methods and appropriate care for patients with Pierre Robin sequence. Databases such as PubMed and Google Scholar were used. Relevant articles were identified using keywords. **Results:** Actions that support the functioning of newborns with Pierre Robin sequence include nasopharyngeal intubation, prone positioning, nasogastric tube feeding, and feeding-facilitating techniques. The first two methods enable airway clearance in the patient. Another challenge in children with Pierre Robin sequence is meeting their caloric needs. Nasogastric tube feeding and a hypercaloric diet may be necessary to fulfil those needs. Nutritive and non-nutritive sucking training also proves helpful. **Conclusion:** Although the treatment of children with Pierre Robin sequence is lengthy, their hospitalisation time can be reduced. Caregivers should accompany patients throughout the hospitalisation period to acquire knowledge and skills that ensure continuity of care after the infant's discharge.

Keywords: Pierre Robin sequence, infant care, cleft palate, glossoptosis, retromicrognathia

Streszczenie

Wstęp: Sekwencja Pierre'a Robina (*Pierre Robin sequence*, PRS) to wrodzone zaburzenie rozwojowe, którego etiologia nie jest do końca poznana. Częstość występowania sekwencji Pierre'a Robina wynosi od 1:8500 do 1:20 000 urodzeń, a śmiertelność – od 3,6% do 21%. Anatomiczne malformacje występujące u pacjentów z sekwencją Pierre'a Robina powodują niedrożność dróg oddechowych i trudności w karmieniu noworodka. **Cel pracy:** Praca powstała w celu podsumowania aktualnego stanu wiedzy na temat nieinwazyjnych metod leczenia i odpowiedniej pielęgnacji pacjentów z sekwencją Pierre'a Robina. **Materiał i metody:** Używając baz danych takich jak PubMed i Google Scholar, autorzy dokonali przeglądu literatury dotyczącej zachowawczych metod leczenia i pielęgnacji pacjentów z sekwencją Pierre'a Robina. W celu znalezienia artykułów posługiwali się słowami kluczami. **Wyniki:** Czynności, które wspomagają funkcjonowanie noworodka z sekwencją Pierre'a Robina, obejmują intubację nosowo-gardłową, ułożenie w pozycji odwróconej na brzuchu, karmienie przez sondę nosowo-żołądkową i techniki ułatwiające karmienie; pierwsze dwie metody umożliwiają udrożnienie dróg oddechowych pacjenta. Innym problemem u dzieci z sekwencją Pierre'a Robina jest zapewnienie właściwej podaży kalorii, co może wymagać zastosowania zgłębnika nosowo-żołądkowego i diety hiperkalorycznej. Pomocny okazuje się również trening ssania odżywczego i nieodżywczego. **Wnioski:** Leczenie dzieci z sekwencją Pierre'a Robina jest długotrwałe, można natomiast skrócić czas pobytu w szpitalu. W tym celu opiekunowie powinni towarzyszyć pacjentom przez cały okres hospitalizacji, aby zdobyć wiedzę i umiejętności, dzięki którym zapewnią ciągłość opieki po wypisaniu niemowlęcia ze szpitala.

Słowa kluczowe: sekwencja Pierre'a Robina, opieka noworodka, rozszczep podniebienia, glossoptoza, retromikrognacja

INTRODUCTION

Pierre Robin sequence (PRS) is a congenital defect arising from first branchial arch malformation[1], which may develop isolated or in association with other syndromes or defects[2]. More than 50 syndromes with co-occurring PRS have been described in the literature[3]. Both genetic and environmental factors are cited as the cause of the defect, while the aetiology of the syndrome is not fully understood. The prevalence of PRS ranges from 1:8,500 to 1:20,000 live births, with mortality estimated between 3.6% and 21%[4,5].

The symptoms of PRS, which include micrognathia, glossoptosis and cleft palate, have a significant impact on a child's development[6]. They often cause airway obstruction and difficulties in feeding the newborn, resulting in delayed weight gain in the first weeks after birth. Due to facial malformation, there is also a high risk of aspiration, which can cause aspiration pneumonia[7].

Therefore, PRS patients require specialised airway management and special positioning to reduce the risk of aspiration and minimise the incidence of the above complications.

Measures to support the functioning of PRS newborns include nasopharyngeal intubation, nasogastric tube feeding, feeding-facilitating techniques, and prone positioning[2].

AIM

The aim of this study was to summarise the current state of knowledge on non-invasive treatment methods and appropriate care for patients with PRS.

MATERIALS AND METHODS

Using PubMed and Google Scholar databases, we reviewed the literature on non-invasive treatment and care for patients with PRS. We used the following keywords to identify relevant articles: "Pierre Robin sequence", "glossoptosis", "retromicrognathia", "infant care", "cleft palate".

RESULTS

Nasopharyngeal intubation

Nasopharyngeal intubation is a recommended method to facilitate the newborn's breathing. Inserting a soft silicone cannula through the infant's nose allows for clearing the airways[2]. The tube should have a diameter of 3.0–3.5 mm. Smaller tubes do not ensure good ventilation, while larger tubes cause great discomfort to the infant. In the hospital setting, nasopharyngeal intubation is performed by nursing personnel, and should be taught to the children's parents or caretakers so that they are able to care for their child after discharge[8].

It is important to check the patency of the cannula after each insertion. This can be done with a strip of gauze applied to the lumen of the tube – the movement of the strip during inhalation and exhalation indicates proper airflow. Then the cannula can be attached to the nasal bridge[8]. The tube should not be removed for the first 24 hours to facilitate the widening of the nostril and the infant's adaptation to the new way of breathing. After 24 hours, the cannula is removed and rinsed under running water[8]. Then, systematic rinsing of the tube is carried out every 6–8 hours. It should be remembered to check the patency of the airway using the method described above after each insertion. Additionally, the child's caregivers should monitor the amount of nasal and oral secretions and suction their excess if necessary[9]. The cannula should be replaced every 7 days as it may lose elasticity under the influence of saliva[8]. After hospital stay, the patient should have 24-hour follow-up visits to the hospital every 15 days to assess whether he or she is able to breathe properly without intubation. Finally, nasopharyngeal intubation can be discontinued when, in the absence of cannula, the saturation is >90% for 24 hours, without deterioration of the child's general condition or the onset of dyspnoea[2].

Prone positioning

Placing a PRS child in the prone position, which allows for clearing the airway in up to 41–69% of patients by naturally helping bring the tongue forward so it does not fall back into the lower pharynx, is an important part of care[10]. Additionally, this position helps evacuate saliva, which often accumulates in excess due to swallowing disorders. This prevents the child from choking or even suffocating. It is also necessary to ensure that the patient lies at a 45-degree angle, which will help improve airway patency and support the treatment of gastroesophageal reflux, which is often seen in children with PRS[2,11]. This positioning should be used not only when lying passively in the crib, but also during baths or basic hygiene and care activities. It is important that medical personnel properly instruct caregivers on how to easily and safely place the infant in the prone position at a 45-degree angle after discharge. For this purpose, pillows or wedges can be placed under the mattress at the headboard of the bed or anti-reflective wedges can be placed against the headboard. It should be also remembered to protect the baby from slipping (e.g., using fabric slings), which makes it easier to maintain the correct position[9].

There are no reliable scientific reports on how long the prone position should be used. Côté et al. shared their own experience, showing that it can take up to several months before there is sufficient improvement in airway patency to place the infant in the supine position. They base their observations on continuous home-based nocturnal SpO_2 monitoring in infants at home[5].

Cole et al. recommend placing the baby on the side instead of prone positioning to reduce the risk of sudden infant death syndrome (SIDS). However, their paper does not present any objective evidence that the proposed position effectively ensures airway patency[12].

Nasogastric tube (NG tube)

Children with PRS have an increased risk of aspiration during feeding. As a result, oral feeding is often contraindicated[2,11]. A nasogastric tube may be necessary to meet energy requirements and reduce the risk of aspiration.

The caregivers should be trained on how to prepare food, feed the child and manage the tube while still in the hospital, shortly after the infant is born[9]. It is important to remember that the baby should be fed in the supine position, with the head elevated by at least 30 degrees[13], and the food introduced should be at room temperature. The tube should be kept clean by rinsing it with water after each feeding to maintain patency[9]. The tube can be removed when the patient is able to take orally 70% of the age- and weight-appropriate portion in less than 30 minutes and without choking, cyanosis or cough[2]. Increasing the portions of food taken orally by the child is possible through feeding training.

Techniques to facilitate feeding

Feeding training tailored to the anatomy of the PRS patient can be introduced under the guidance of a speech therapist. In some cases, oral feeding of the child is already possible after about 7 days of treatment[13,14]. Non-nutritive and nutritive sucking training may be distinguished. Non-nutritive sucking training is performed using either relaxing massage with the tongue extended forward or manual mandibular support during sucking. During nutritional sucking, the patient's caregivers should pay attention to the way the tongue is placed on the bottle teat and whether the movements are rhythmic. Additionally, it is advisable to use a long and soft bottle teat with an opening enlarged to 1 mm to facilitate food intake[2,11]. The important role of a hypercaloric diet has also been described. It promotes the overall growth and development of the child and, as a result, early removal of the gastrointestinal tube[2].

Palatal plates

Reports on the use of orthodontic appliances (palatal plates) in the treatment of patients with PRS may be also found in the literature. However, these publications are too scarce to objectively confirm the efficacy of such methods[5]. The same is true for a tool known as the pre-epiglottic baton plate (PEBP). By shifting the tongue into a more anterior position, it widens the lower pharyngeal space. Preliminary studies have shown that PEBP reduces airway obstruction; however, larger, multicentre studies to ensure reproducibility of the results are missing[5].

CONCLUSIONS

Due to the risk of aspiration and airway obstruction, patients with PRS require special care from birth, both from medical personnel and their caregivers. Although PRS children require long-term conservative treatment, there is no need for them to stay in the hospital all the time. Caregivers should accompany the infant throughout hospitalisation to gain knowledge and skills to ensure continuity of care after hospital discharge[15]. It is worth mentioning that the presented conservative treatment methods are particularly intended for patients before surgical correction of the defect and infants. Significant functional improvements can be seen around the age of 1 year. As the child grows, the mandible widens and the coordination of the tongue and adductor muscles improves. This leads to better anatomical conditions translating into reduced airway obstruction and improved child feeding[16].

Conflict of interest

The authors report no financial or personal relationships with other individuals or organisations that could adversely affect the content of the publication and claim ownership of this publication.

Author contributions

Original concept of study: NG, PR, TZ, AN, AR, OJ, AM. Collection, recording and/or compilation of data: NG, PR, TZ, AN, AR, OJ, AM. Analysis and interpretation of data: NG, PR, TZ, AN, AR, OJ, AM. Writing of manuscript: NG, PR, TZ, AN, AR, OJ, AM. Critical review of manuscript: NG, PR, TZ, AN, AR, OJ, AM. Final approval of manuscript: NG, PR, TZ, AN, AR, OJ, AM.

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