RESEARCH PAPER | Medical Science | Volume : 6 | Issue : 6 | June 2016 | ISSN - 2249-555X | IF : 3.919 | IC Value : 74.50

Pierre Robin Syndrome-A Review

KEYWORDS: Pierre Robin, Cleft palate, sleep apnea, airway obstruction

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ABSTRACT *Pierre Robin Syndrome is also known as Pierre Robin Sequence (PRS). The features of this syndrome are small mandible (micrognathia), glossoptosis and U-shaped cleft palate. It is a rare malformating pathology and its abnormality happen among infants. Pierre Robin Sequence may be caused by genetic anomalies at chromosomes 2, 11 or 17. The goals of treatment in infants with Robin Sequence focus upon breathing and feeding, optimizing growth and nutrition despite the predisposition for breathing difficulties. There are several treatments can be done such as nasopharyngeal cannulation. Several case reports are seen in the babies which have the features of Pierre Robin Syndrome.*

Introduction

Pierre Robin Syndrome (PRS) was first introduced as glossoptosis by Pierre Robin, a French Physician in 1923 [1]. This syndrome is associated with micronagthia, glossoptosis, airway obstruction and wide U-shaped cleft palate which is reported to be 90% of the cases. However, this syndrome is uncommon among infants which have ratio of 1: 5000 to 1: 85000. One half of the cases are mostly syndromic which is more prevalent to high mortality. The common syndrome are Stickler, velocardiofacial and Treacher-Collins[2]. Airway obstruction and feeding are the problems faced by the infants with PRS. If it is left untreated, it may lead to acute and chronic hypoxia, apnea, cyanosis, aspirations, respiratory tract infections, malnutrition and failure to thrive [3]:B.

Recently, it is found that genetic anomalies is one of the causes of PRS especially at the chromosome 2, 11 and 17 [4] Non-syndromic PRS may be caused by SOX9 and KCNJ2 dysregulation [5]. Infant with PRS family member has a higher probability of cleft palate.

PRS can be solved and treated by variety of treatments. The treatment for PRS can be either surgical or non-surgical interventions. Non-surgical is the most preferable method and 68% of the cases were successfully treated. However, surgical method are still being used such as tracheostomy and tongue lip adhesion method. Several case reports with different diagnosis and treatment were also reviewed.

Materials and method

50 articles were selected based on the topic of this article and have been reviewed. The method and discussion of each research article have been thoroughly read. Therefore, during the review session, 19 articles have been selected and the current knowledge about the article has been reviewed.

Discussion

Pierre Robin Syndrome (PRS) is associated with airway obstruction and feeding difficulty. Therefore, several methods were proven to help reduce the problems either through surgical or non-surgical method.

Non-surgical method

Prone or lateral positioning

Generally, prone positioning is the most preferable and simplest management to recover the airway obstruction in infants with PRS. 70% of the cases has been solved through this method [6]. However, Daniel. M et al [7] have done a study which shows that infant with PRS mostly had obstructive sleep apnea even though prone positioning is an effective method in relieving airway obstruction.

Therefore, lateral or side positioning is recommended by Cole et al as prone positioning is related with Sudden Infant Death Syndrome (SIDS)[8].

Nasopharyngeal airway

If prone positioning seems difficult to reduce the airway obstruction, nasopharyngeal airways (NPA) can be done by placing endotracheal tube intranasally and positioned in the distal oropharynx, beyond the area of obstruction by glossoptosis [4]. Polysomnography is used to determine the timing of removal of (NPA). It can be done by the parents itself at home. However, proper technique should be taught to obtain a good result.

Orthodontic apparatus

Some centres use orthodontic apparatus to treat feeding difficulties instead of to solve airway obstruction. Recently, pre-epiglottic baton plate (PEBP) was introduced to help reduce problem of Obstructive Sleep Apnea (OSA) among infants. It is done by shifting the tongue anteriorly and thus widen the hypopharyngeal space [4]. PEBP is customized for every infant with PRS. Cast is taken and send to the laboratory for manufacturing. The PEBP is made up of compound soft and hard acrylic.

Surgical method

Surgical method would be the second option after non-surgical method. If the infants with PRS are facing severe or moderate airway obstruction and the non-surgical method such as nasopharyngeal tube, laryngeal mask does not give any good result, it would be best to perform surgical method such as tongue lip adhesion, mandibular distraction osteogenesis and last option would be tracheostomy.

Tongue lip adhesion (TLA)

In 1911, tongue lip adhesion method was introduced and expressed in detail by Shukowsky [9]. However, around 20th century, Douglas took an initiative to popularize this method as it is the most efficient way to reduce airway obstruction in infants with PRS in certain circumstances.

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Tongue lip adhesion would be best done in infants who have Stickler syndrome and velocardiofacial syndrome due to good prognosis of mandibular growth [10].

The procedure is done by adhering the base of the tongue to the lower lip through suturing. This procedure opens the oropharyngeal airway space as the tongue base is pulled forward [3]. TLA is advised to be done in infants who have not start the development of the teeth. This is because it may interrupt the process and reduce the chances of healing.

There are four type of obstructions which are Type I, II, III and IV and it is related with PRS in infants [11]. Type I obstruction is the most common and it can be treated fully by TLA. It is caused by posterior movement of the tongue against posterior pharyngeal wall. Type II obstruction is due to posterior and superior displacement of the tongue, promoting contact between the tongue, the velum and the pharyngeal wall in the superior oropharynx. Type III is a pharyngeal obstruction caused by prolapse of the medial pharyngeal wall. Type IV is due to constriction of the pharynx in a circular manner by movement of the tongue and the lateral pharyngeal walls [3]¤ . However, Type II, III and IV is not suitably done by TLA.

If there is failure in TLA, other method can be done as initiative such as mandibular distraction osteogenesis (MDO) or tracheotomy.

Mandibular distraction osteogenesis (MDO)
Mandibular distraction osteogenesis is introduced by McCarthy in year 1989. It plays an important role in treating infants with PRS and correction of deformity with minimal morbidity [12]. Besides, it is popular not only for treating airway obstruction in PRS, but it also improve the facial cosmetics and malocclusion.

However, it is believed that MDO procedure lead to several complication on the infants such as damaged tooth buds, inferior alveolar nerve injury and unsightly facial scars [3]. Besides, pin site infections, unacceptable scarring, resorption/ankylosis at the temporomandibular joint, malunion, and failure of distraction due to incomplete osteotomies, early consolidation, or device failure are also the complications that may be faced by the patient [13, 14]. Therefore, it can only be do in the minority of infants.

Apart from that, MDO can lead to mandibular prognathism if the infants have a normal relation of lower jaw position as in velocardiofacial syndrome and Stickler syndrome[15]□(.

The obstruction in airway of the infants can be relieve through osteotomy which helps in advancing the mandible. Thus, the supraglottic airway obstruction can be reduced by bringing the tongue base forward[16]. It also indirectly correct the micrognathia and glossoptosis.

The advantages of MDO is it can be the alternative way to avoid tracheotomy. In some cases, the patient is successfully decannulated after the MDO was completed and the patient is underwent tracheotomy prior to MDO [17].

Tracheostomy
Tracheostomy is one of the procedure that can be done to treat PRS in infants. It can be done with local anaesthesia or general anaesthesia. However, this method is currently would be the last choice among the other method. Although, a survey stated that tracheostomy to be the safest and most reliable method for a long term airway management for the infant with PRS, there are several complications related to it [18]. For example, tracheal stenosis, granuloma formation, tracheal fistula formation, cannula obstruction and accidental decannulation [18]. 19% to 49% of patients have been reported to diagnose with this complication[18].

One of the consequences of tracheostomy is the infants who was treated initially with tracheostomy may shows a significant late in the production of speech and development of language.

Case report review
Krishna et al reported a case of a neonate with PRS. The infant experienced an airway obstruction and respiratory distress after birth due to acute glossoptosis and microretrognathia. Initially, non-surgical innervation was done such as using nasal cannula and prone positioning. Then, TLA was done under general anaesthesia due to poor weight gain. The infant shows improvements after the procedure. For example, he starts to gain weight, reduced episodes of respiratory infection and improvement in oxygen saturation. After one year, the suture was removed and the infant was completely stable without respiratory distress.

In another cases, Onal et al shared an experience dealing with PRS neonates. The patient has difficulty to be intubated and he had a history of tracheostomy. He has difficulty in mouth opening, micrognathia, short extremities, mild airway obstruction and respiratory distress[19].

Conclusion
There are variety of procedure in managing PRS in infants including difficulty in airway obstruction and feeding. It can be done by reviewing the diagnosis as it helps in managing the problems effectively.

This review is focusing on the treatment of PRS and few cases which reported on PRS in infants.

References

1. Gupta, R., M. Patel, and N. Bajaj, *Pierre Robin syndrome: a case report.* International Journal, 2015. **3**(11): p. 3432.
2. Sesenna, E., et al., *Mandibular distraction in neonates: indications, technique, results.* Ital J Pediatr, 2012. **38**(7).
3. Krishna Kumar, K., et al., *Tongue-lip adhesion in Pierre Robin sequence.* Journal of the Korean Association of Oral and Maxillofacial Surgeons, 2016. **42**(1): p. 47-50.
4. Andrews, S., et al., *Surgical management of a large cleft palate in a Pierre Robin sequence: A case report and review of literature.* J Pharm Bioallied Sci, 2015. **7**(Suppl 2): p. S718-20.
5. Agrawal, K., *Cleft palate repair and variations.* Indian Journal of Plastic Surgery, 2009. **42**(3): p. 102.
6. Anderson, I.C.W., et al., *Prevalence and severity of obstructive sleep apnea and snoring in infants with Pierre Robin sequence.* The Cleft Palate-Craniofacial Journal, 2011. **48**(5): p. 614-618.
7. Daniel, M., et al., *Airway, feeding and growth in infants with Robin sequence and sleep apnoea.* International journal of pediatric otorhinolaryngology, 2013. **77**(4): p. 499-503.
8. Dwyer, T. and A.-L. Ponsonby, *Sudden infant death syndrome and prone sleeping position.* Annals of epidemiology, 2009. **19**(4): p. 245-249.
9. Rogers, G.F., et al., *The GILLS score: part I. Patient selection for tongue-lip adhesion in Robin sequence.* Plastic and reconstructive surgery, 2011. **128**(1): p. 243-251.
10. Abramowicz, S., et al., *Validation of the GILLS score for tongue–lip adhesion in Robin sequence patients.* Journal of Craniofacial Surgery, 2012. **23**(2): p. 382-386.

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11. Sher, A.E., R.J. Shprintzen, and M.J. Thorpy, *Endoscopic observations of obstructive sleep apnea in children with anomalous upper airways: predictive and therapeutic value.* International journal of pediatric otorhinolaryngology, 1986. **11**(2): p. 135-146.
12. Master, D.L., P.R. Hanson, and A.K. Gosain, *Complications of mandibular distraction osteogenesis.* Journal of Craniofacial Surgery, 2010. **21**(5): p. 1565-1570.
13. Fritz, M.A. and J.D. Sidman, *Distraction osteogenesis of the mandible.* Current opinion in otolaryngology & head and neck surgery, 2004. **12**(6): p. 513-518.
14. Denny, A. and C. Amm, *New technique for airway correction in neonates with severe Pierre Robin sequence.* The Journal of pediatrics, 2005. **147**(1): p. 97-101.
15. Rogers, G., et al., *Effect of a syndromic diagnosis on mandibular size and sagittal position in Robin sequence.* Journal of Oral and Maxillofacial Surgery, 2009. **67**(11): p. 2323-2331.
16. Côté, A., et al., *Pierre Robin sequence: Review of diagnostic and treatment challenges.* International journal of pediatric otorhinolaryngology, 2015. **79**(4): p. 451-464.
17. Boston, M. and M.J. Rutter, *Current airway management in craniofacial anomalies.* Current opinion in otolaryngology & head and neck surgery, 2003. **11**(6): p. 428-432.
18. Kirschner, R.E., et al., *Surgical airway management in Pierre Robin sequence: is there a role for tongue-lip adhesion?* The Cleft palate-craniofacial journal, 2003. **40**(1): p. 13-18.
19. Onal, O., et al., *Our Experience With Regional Anesthesia in a Case of Pierre Robin Syndrome.* Anesthesiology and Pain Medicine, 2016. **6**(1).