Case Report
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Pierre robin sequence: case report and review

Summary

Pierre Robin sequence is a condition where baby is born with a small lower jaw, is typically described as micrognathia, glossoptosis, and cleft palate. Pierre Robin sequence can be an isolated finding, or can be associated with a genetic syndrome in approximately 50% of cases.

1.1. Objective: Report a Pierre Robin sequence in a neonate at Caracas Universitary Hospital.

1.2. Clinical case: Full-term male neonate shows signs of gastroesophageal reflux during breastfeeding and alteration in the palate, which is why they request an evaluation by the Plastic and Reconstructive surgery service. At the physical examination, adequate mouth opening is evident: a palatal cleft is visualized that involves the hard and soft palate of approximately 25 x 30 mm, which allows visualization of the posterior surface of the pharynx. Absence of choanae with hypoplasic tongue. Suction reflex preserved with decreased pressure. Findings are complete palatal cleft, microglossia and micrognathia.

1.3. Discusion and conclusion: Clinicians mainly focus on the management of the morbidities caused due to PRS. Managing respiratory obstruction is prioritized over addressing feeding dysfunction, however; our patient does not have this problem. Supplemental feeding using a nasogastric/orogastric tube (mild cases) and gastronomy tube (chronic and persistent cases) helps resolving feeding difficulties. Despite the relative low frequency, the Pierre Robin sequence is a complex scenario for any neonate and his family. Requires a multidisciplinary team in order to obtain the best results and outcomes avoiding complications.

Keywords: pierre robin sequence, clef palate, micrognathia, neonate

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Introduction

Pierre Robin sequence, first described in the French literature in 1934, is a condition where baby is born with a small lower jaw, is typically described as micrognathia, glossoptosis, and cleft palate.[1] Other authors classically defined as micrognathia, glossoptosis, and upper airway obstruction, with or without a cleft palate. This sequence frequently contributes to feeding problems which may delay oral feeding.[2] The incidence reported in the literature ranges from 1/31206 live births to 1/8060 live births. Pierre Robin sequence can be an isolated finding, or can be associated with a genetic syndrome in approximately 50% of cases.[3]

Case report

Full-term male neonate, 48 hours of birth product of II gestation of pregnancy of 39 weeks with negative amniocentesis and controlled pregnancy, obtained by simple eutocic delivery where the neonatology service shows signs of gastroesophageal reflux during breastfeeding and alteration in the palate, which is why they request an evaluation by the Plastic and Reconstructive surgery service. At the physical examination, adequate mouth opening is evident: a palatal cleft is visualized that involves the hard and soft palate of approximately 25 x 30 mm, which allows visualization of the posterior surface of the pharynx. Absence of choanae with hypoplasic tongue. Suction reflex preserved with decreased pressure. Round nasal tip is evident. Findings are complete palatal cleft, microglossia and micrognathia which are compatible with the Pierre Robin sequence diagnosis. Work plan begans by evaluation of Pediatric dentistry (consider palate plaque) and evaluation by phoniatrics and performing exercises to stimulate the muscles of the soft palate while it is assessed to be able to the future surgical treatment.

Discussion

Considering the pathogenesis of the disease, three major theories have been proposed: mechanical theory, neurological maturation theory, and mandible compression theory. It is important to distinguish between non-sindromatic PRS and sindromatic PRS as the treatment strategy in the latter would have to account for PRS and the associated syndrome.[4] In the presented case the rest of the physical examination shows a NS PRS Figure 1&2.

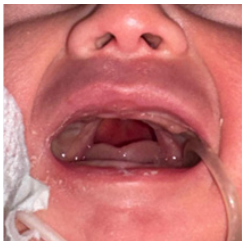

Figure 1 Clef palate in a neonate with PRS.

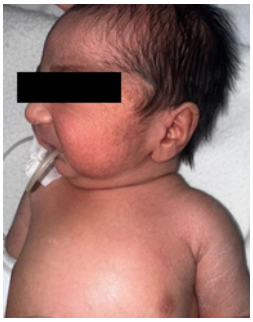

Figure 2 Micrognatia is evident in a neonate with PRS.

Cleft palate is frequently encountered, but is not considered as a prerequisite for the diagnosis,[5] having relation with our case because the cleft palate was the initial finding causing the gastroesophageal symptoms. Ultrasound imaging helps in the prenatal diagnosis of severe cases of Pierre Robin sequence and helps specialists plan the further course of pregnancy and postnatal care.[6] However in the patient was not possible to recognize these findings during the pregnancy. When the triad of PRS is spotted, the physician should have an heightened index of suspicion for other anomalies and a detailed family history of congenital anomalies should be elucidated.[7] Follow these words the mother had not any both familiar and personal antecedents of congenital anomalies.

Clinicians mainly focus on the management of the morbidities caused due to PRS.[8] Managing respiratory obstruction is prioritized over addressing feeding dysfunction,[9] however; our patient does not have this problem. Supplemental feeding using a nasogastric/orogastric tube (mild cases) and gastronomy tube (chronic and persistent cases) helps resolving feeding difficulties.[10] However in our case by the guide of our Specialist it recommends to the neonatology service that they should continue the breastfeeding with special indications in order to help him nutritionally and began the functional orthopedics of the maxilla in order to improve his status and for the future surgical intervention.

Conclusion

Despite the relative low frequency, the Pierre Robin sequence is a complex scenario for any neonate and his family. Requires a multidisciplinary team in order to obtain the best results and outcomes avoiding complications.

Acknowledgments

To the San Juan de Dios Hospital, Plastic and Reconstructive Unit and Neonatal Unit of the Caracas Universitary Hospital

Conflicts of interest

The author declare no conflicts of interest.

Ethical approval

This research complies with the World Medical Association Declaration of Helsinki on medical protocols and ethics.

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