

Identical twins with Pierre Robin syndrome; unusual encounter

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Type of article: Case report**Abstract**

New born babies could suffer from multiple craniofacial abnormalities, such as Pierre Robin syndrome, which consists of micrognathia and relative macroglossia with or without cleft palate. Although Pierre Robin syndrome is well described in literature, only a few have mentioned its occurrence in identical twins. This paper presents a rare incident of full-term twin babies born with the sequence of Pierre Robin syndrome, which consists of micrognathia, cleft palate, and glossoptosis. Although it is a rare coincidence, Pierre Robin syndrome still can occur in identical twin babies. The treatment is a step-by-step approach, but all procedures are mainly directed to widening the pharyngeal space.

Keywords: Cleft palate, Macroglossia, Micrognathia, Pierre Robin**1. Introduction**

Although Pierre Robin described this clinical entity in 1923 and then described the full triad to include cleft palate in 1934 (1), the clinical components of this sequence were described by Von Siebold in 1835, and it was Fairbairn who mentioned the full triad in 1846 (2). Butow and his colleagues had reviewed the literature and recommended that Pierre Robin syndrome (PRS) should be subdivided into Fairbairn–Robin triad and Siebold–Robin sequence (3). Pierre Robin syndrome is a sequence; its three main features are retrognathia, cleft palate, and glossoptosis. Breathing and feeding difficulties that result from these anomalies might require emergency management (4). The exact cause is not known, but it could be present at birth as a part of many genetic syndromes. External factors that interfere with the growth of the lower jaw may contribute to PRS. Some neurological conditions can decrease the jaw movement in utero restricting its growth, such as Stickler syndrome and Nager syndrome. These patients could be diagnosed before birth with the help of ultrasonography or immediately after birth. They undergo multiple surgeries under general anesthesia. Pierre Robin syndrome incidence in identical twins is rare but still occur. In this paper, we will present a case report of a rare incident of full-term identical twin babies born with the full sequence of Pierre Robin syndrome.

2. Case Presentation**2.1 History**

Full-term identical twin baby boys delivered by cesarean section after noneventful pregnancy. Birth weights were 2,100g and 2,400g. Apgar score at birth was 7/10, and at 5 min was 8/10. Formal consultation was sent to our team in Prince Hashem bin Al-Hussein Military Hospital–Zarqa city/Jordan on 20 July 2016 by pediatrician; neonates were diagnosed to have Pierre Robin syndrome, for further evaluation and management.

2.2. Clinical Examination

Upon extraoral clinical examination, both neonate showed mandibular retrognathia, a characteristic bird-face appearance (Figure 1). Intraoral examination revealed glossoptosis, palatal malformations, and cleft palate. Family history of the patients revealed no similar cases, and there was no history of cleft palate or facial anomalies in the family (Figure 2).

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2.3. Investigations

For both infants, the full blood count and kidney function tests were within the normal range; 2D echo revealed patent foramen ovale, brain ultrasound showed no abnormalities, and renal ultrasound showed no abnormality.



Figure 1. Identical twins presented with PRS.



Figure 2. Photo of neonate at age of six weeks showing cleft palate.

3. Discussion

Pierre Robin syndrome is a craniofacial anomaly comprising mandibular hypoplasia, cleft of the secondary palate and glossoptosis, which might lead to a life-threatening obstructive apnea and feeding difficulties during neonatal period. It is reported that Pierre Robin syndrome has an incidence that ranges between 1/8500 and 1/14,000 births (5, 6). The incidence of twins with PRS support a genetic theory as a cause; also supporting this theory is the higher

incidence of cleft lip and palate in family members (7). Identical twin brothers' cases suffering from PRS are very rare; the literature reported that Sachtleben described two brothers who suffered from PRS symptoms, which were associated with other congenital anomalies (8). Shah et al. also had described PRS in four siblings, which included a set of twins (9). Smith and Stowe reported affected brothers in 1961 (10), and a year later patients were pictured by McKusick who raised the possibility that these brothers might had either Stickler syndrome or Wagner syndrome (11). Others argued that Stickler syndrome should be considered first in cases of the Pierre Robin syndrome, especially in familial incidences (12). Management of infants with PRS is focused mainly with functional issues associated with airway and feeding. Thirty percent mortality has been mention, but the prognosis is good if the neonate survive the respiratory and feeding issues (13). Thus, treatment may be as a surgical emergency in the neonatal period to the symptomatic management of upper respiratory obstruction and feeding problems. All procedures are mainly directed to widening the pharyngeal space or bridging the narrow upper airway; these range from postural positioning to use of appliances to surgical procedures (14). Our plan in management of such cases is a step-by-step approach as follows: On day one, a tongue tie was done with lingual mucosa (glossopexy) to facilitate soft nipple feeding with wide opening or squeeze bottle (15). Palatal feeding obturator temporary constructed for both twins until surgical treatment is initiated to close the cleft palate for the purpose of ease of feeding and to prevent regurgitation or aspiration and to correct tongue positioning and speech development and reduce the incidence of nasopharyngeal infections (16). An 18-month surgical procedure using the von Langenbeck technique, including iliac crest bone graft, will be done to close the palatal cleft (17). At the moment, both babies are growing well, and their mandibular growth is under observation. We are planning an orthodontic approached treatment at the mixed dentition stage between 9 to 11 years to expand the palate and advance the mandible to stimulate its growth (18). Surgical treatment after the child turns 18 years old includes orthognathic surgery and mandibular distraction osteogenesis (19).

5. Conclusions

Although it is a rare coincident Pierre Robin syndrome still can occur in identical twin babies. The treatment is a step-by-step approach, but all procedures are mainly directed to widening the pharyngeal space.

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Conflict of Interest:

There is no conflict of interest to be declared.

Authors' contributions:

NS is responsible for the concept of the case report. NS and KM wrote the manuscript. ZZ, HK, and RO reviewed and edited the manuscript. NS and KM reviewed references. All authors read and approved the final manuscript.

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