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Tongue--tied: Management in pierre robin sequence, a case report

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Pierre Robin Sequence (PRS) is a rare congenital condition of facial abnormalities, defined by a triad of micrognathia, retroglossoptosis and airway obstruction. PRS may have varied presentations due to associations with syndromes. Hence, the consensus in management remains elusive, with no definitive treatment protocols. We describe a case from a resource--scarce setting that highlights the use of a less commonly performed surgical procedure which is simpler and cheaper than the gold--standard surgery in PRS. An 18--month--old boy with PRS presented to A&E with airway obstruction and hypoxia due to retroglossoptosis. He was resuscitated immediately and intubated. Gold--standard treatment was surgery: Mandibular Distraction Osteogenesis (MDO). However, as the patient was unable to afford the surgery, a cheaper procedure called tongue--lip adhesion (TLA) was performed. The procedure involved pulling the base of the tongue anteriorly and tying to the hyoid bone. This maintained airway patency and patient was extubated. Mother was given feeding and child positioning advice. It is expected that the mandibular growth will eventually catch up with the tongue growth. The surgical procedures used to relieve airway obstruction in PRS include TLA, MDO and tracheostomy. In this case, TLA was chosen due to affordability issues. Three different TLA techniques, previously described in the literature for PRS, were discussed. Given the nutritional status of the patient, we decided to avoid extensive dissection. We required a technique that would not restrict mobile segments of the tongue, to allow for normal speech development and feeding. In addition, sutures on the tongue should not be damaged by biting, in the teething child. Lapidot and Ben--Hur technique (briefly described in the case) satisfied above--mentioned requirements and was hence chosen. Overall, this case is of great value in exploring different surgical techniques for PRS management, not widely explained in the literature.

Biography

Nikitha Rajaraman is a 4th year medical student from the University of Glasgow. She is a highly motivated in pursuing her career in the surgical specialities. She has completed modules and electives in General, Plastic, Upper GI and Vascular Surgeries, and has completed various audit projects in the process. She has also contributed to national audits. She was recently awarded the top poster prize at the 8th Surgical Undergraduate Conference 2017 conducted in the Royal College of Physicians and Surgeons of Glasgow. She was also awarded Senior Elective award by the university as a recognition for her efforts towards a surgical career.

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