

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

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Abstract

Introduction: 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.

Background: 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.

Method:- PRS is described by an exemplary group of three of micrognathia, glossoptosis, and aviation route block. Microretrognathia is quickly recognized during childbirth and is a characterizing highlight of the determination. Hypoplastic mandibles are little in both the vertical and flat measurements. This hence represents the abatement in the anteroposterior projection of the jaw and its ensuing great retrognathic appearance. Aside from the micrognathia, Randall portrayed the prominent finding of retrogenia, or

back uprooting of the jaw, to describe the starting inconsistency in this grouping. Glossoptosis, characterized as an anomalous back position of the tongue, is the subsequent trademark highlight of PRS. The situation of the tongue is to a great extent dictated by the size and direction of the mandible. As a littler mandible has less front projection, the tongue will thus be moved posteriorly. In addition, despite the fact that the tongue is ordinarily noted to be of typical size, the hypoplastic mandible gives less volume in the oral pit and powers the tongue to fit into a littler space, which further serves to fuel the blockage of the back pharynx.

Results: The treatment of newborn children with PRS stays dubious and contrasts among establishments. Our way to deal with the newborn child with PRS starts with endeavors at inclined situating to take out the impact of gravity on the base of the tongue. Nonstop heartbeat oximetry is performed on all patients. In the event that situating alone isn't effective, adjunctive estimates, for example, supplemental oxygen and adjusted nasopharyngeal cylinders are utilized to sidestep the tongue base deterrent. We find that polysomnography can be a valuable device to record the obstructive apnea--hypopnea file and affirm the nonattendance of unrecognized obstructive or focal rest apnea. Another key element of our treatment centers around taking care of and dietary help. All patients are assessed intently with respect to their capacity to take care of by qualified language instructors who have practical experience in taking care of. In the event that essential, early nasogastric taking care of is started to enhance oral feeds and improve weight gain. Most newborn children will be effectively dealt with moderate estimates alone. In the event that these measures neglect to ease block as confirm by deficient outcomes on rest studies and helpless weight increase, careful alternatives are thought of. Prior to any surgical intervention, we confirm the absence of obstruction below the level of the tongue base with direct laryngoscopy and bronchoscopy performed by a pediatric otolaryngologist. Though numerous algorithms are described in the literature in regards to surgical decision making, no uniform consensus currently exists. The clear benefits of one surgical technique versus another have not clearly been determined and neither procedure can be expected to provide normal occlusion for individuals at the time of skeletal maturity.

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,

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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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