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Pediatric anesthesiologists are frequently consulted to formulate a perioperative plan for neonates who present with medical challenges within the first seconds of life. In this case report, we describe the approach to airway management of a neonate with Larsen Syndrome (LS) immediately after caesarean delivery. LS is a rare genetic disorder caused by a defect encoding filamin B, a protein involved in cytoskeletal regulation. Patients with this congenital disorder may present with cardiac defects, tracheomalacia, dysmorphic facial appearances, limb contractures, and other anomalies.

A 22 year-old parturient with LS presented to the labor and delivery suite. Her birth history was significant for tracheomalacia with a difficult intubation requiring a tracheostomy at birth. During her pregnancy, prenatal ultrasound and magnetic resonance imaging detected fetal anomalies suggestive of LS. The pediatric anesthesiology and otolaryngology surgery teams were consulted for airway management of the neonate after a scheduled cesarean delivery.

The pediatric anesthesiology service coordinated the operating room logistics for this case. Two connecting operating rooms would be needed to care for mother and neonate. Once the baby is delivered, the neonatal intensive care unit (NICU) team would carry the baby to the neonate's operating room and perform their initial survey and resuscitation. The neonate's operating room would have a difficult airway cart including a pediatric GlideScope®, rigid bronchoscope, laryngeal mask airways, multiple oral endotracheal tube sizes and an open tracheostomy set ready for use.

Three scenarios were devised to encompass all delivery outcomes. Plan A described the neonate with adequate Apgar scores, breathing appropriately, and not requiring supplemental oxygen. No airway intervention would be indicated. Plan B described the neonate experiencing respiratory distress requiring supplemental oxygen. Direct laryngoscopy would be performed and the airway would be secured with an oral endotracheal tube. Plan C involved neonatal respiratory distress along with a failed direct laryngoscopy. A rigid bronchoscopy would be performed with tracheostomy as backup.

Our neonate was delivered and the NICU team performed the initial assessment of the baby while a peripheral intravenous line was obtained and standard monitors were placed. The patient was noted to have respiratory distress with sternal retractions. An awake direct laryngoscopy was performed and a Cormack and Lehane Grade 1 view was obtained with notice of a bifid uvula and tracheomalacia. A 2.5 mm uncuffed oral endotracheal tube was inserted with no air leak noted around the tube.

In our case, preparation of the operating room in anticipation of a difficult neonatal airway and communication amongst the surgical, obstetric, critical care, and anesthesia teams was essential. After intubation, the neonate was transported to the NICU for further medical treatment.



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