Use of the LMA Classic to Secure the Airway of a Premature Neonate with Smith-Lemli-Opitz Syndrome: A Case Report

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In addition to managing the most challenging of airways within the operating room environment, anesthesia providers are frequently consulted or requested to participate in emergency airway control in various areas of the hospital, often after other providers have failed.

The following is a case report of a premature infant born with multiple and life threatening congenital anomalies in a rural facility. The current recommendations of the American Heart Association for neonatal resuscitation were followed; however, the resuscitating team was unable to secure the airway using standard intubating techniques. Consultation with the anesthetist on duty resulted in the successful placement of the laryngeal mask airway (LMA) size 1.

The pediatrician involved in the care of the patient had minimal experience with using the LMA; however, with verbal instruction was able to successfully place the LMA. With a patent airway established, the patient stabilized and was transferred to a tertiary facility for aggressive care. Although currently not part of the American Heart Association neonatal resuscitation algorithm, consideration of the LMA as a tool to manage an airway after failed attempts at intubation may be appropriate.

Key words: Laryngeal mask airway, neonate, Smith-Lemli-Opitz syndrome.

Smith-Lemli-Opitz syndrome is a multiple congenital anomaly syndrome involving facial and orolaryngeal defects with principal airway implications characteristic of cleft palate, microcephaly, and micrognathia, which may be associated with Pierre Robin syndrome. Bag mask ventilation and intubation may be difficult. The literature suggests that experience with the use of the laryngeal mask airway (LMA) in neonates is emerging and supports the use of this device in the neonatal population. Data on the employment of this airway adjunct in the premature neonate also is slowly emerging. The following is a case report detailing the successful use of this tool to manage the airway of a premature neonate with Smith-Lemli-Opitz syndrome that was born with craniofacial and orolaryngeal birth defects in a rural facility.

Background
Smith-Lemli-Opitz syndrome (SLOS) was initially described in 1964. The syndrome has been known by many names, which has led to confusion in this regard. The accepted terms are “SLOS type I” and “SLOS type II.” SLOS is characterized by variable developmental delay, dysmorphic facial features, micrognathia, cleft palate, and a host of anomalies affecting the central nervous, cardiac, respiratory, gastrointestinal, and genitourinary systems. SLOS is an inborn error of metabolism that includes the accumulation of a toxic precursor of cholesterol (7-dehydrocholesterol) and a deficiency of cholesterol (Figure). Affected individuals usually have a low plasma cholesterol level and elevated levels of cholesterol precursors. Severely affected individuals have multiple congenital anomalies (Table 1) and are often miscarried, stillborn, or die within the first few weeks of life (SLOS type II). Dysmorphic facial features and mental retardation are typical. Mildly affected individuals may have only subtle dysmorphic features with learning and behavioral disabilities (SLOS type I).³

The incidence of SLOS is estimated to be 1:20,000 to 1:60,000 among whites. Its incidence among other races has not been studied; however, it has been described in the United States, northern Europe, Japan, South America, and other countries. The carrier frequency is 1:30 in those of northern European descent suggesting a disease incidence between 1:5,000 and 1:18,000.⁴ The actual disease incidence is less due to fetal losses. The syndrome is heterogeneous with an X-linked Opitz syndrome (XLOS) form and has previously been described as an autosomal recessive disorder; however, it has been recently reported as an autosomal dominant Opitz syndrome (ADOS) form.⁵

Case summary
A 20-year-old primigravida patient arrived in the emergency room with complaints of low back pain. Examination of the patient revealed a 36-week pregnancy, breech presentation, and a cervix that was fully...
dilated. Although this rural facility does not routinely provide obstetric care, the patient's progress in labor necessitated that she be admitted to the hospital for cesarean delivery. The neonatal team from the closest tertiary care facility and a local pediatrician were summoned to be available for the delivery.

Upon delivery the neonate was observed to be hypotonic with little respiratory effort. Appropriate American Heart Association resuscitative steps were initiated (drying, warming, and bag mask ventilation) with poor response. Cardiopulmonary resuscitation was initiated and continued for 60 seconds when a heart rate of 100 beats per minute was noted. The patient was observed to take weak ineffective breaths, therefore, bag mask ventilations continued with auscultation of diminished breath sounds. The Apgar score was 0 at 1 minute and 2 at 5 minutes.

The team transferred the patient out of the operating room to continue with aggressive care and resuscitation. The pediatrician decided to intubate the patient. The neonate was observed to have multiple congenital anomalies. A high arched palate, cleft tongue, micrognathia, broad nasal tip with anteverted nostrils, and low set ears were immediately apparent. Multiple intubation attempts by the pediatrician and resuscitating team proved futile, as visualization of the vocal cords was difficult. Resumption of bag mask ventilation afforded fair response. An umbilical line was placed and arterial blood gases (ABGs) were drawn. The results of the ABGs illustrate that the patient was oxygenated; however, he was not adequately ventilated as demonstrated by the increasing PCO$_2$ (respiratory acidosis) (Table 2).

Consultation with the hospital anesthesia provider concluded with the recommendation to attempt placement of the LMA Classic size 1 (patient weight: 2.3 kg). The anesthesia provider, who was the sole anesthesia provider available to the facility and currently conducting an anesthetic on another patient, coached the pediatrician through the technique for placing the LMA. There is limited data on the use of the LMA in the premature neonate, so the pediatrician

Table 1. Congenital anomalies that may appear in patient with Smith-Lemi-Opitz syndrome

- Microcephaly
- Broad nasal tip with anteverted nostrils
- Micrognathia
- Ptosis of eyelids
- Epicanthal folds
- Strabismus
- Cataracts
- Broad maxillary alveolar ridges
- Slanted or low set ears
- Syndactyly of second and third toes
- Postaxial polydactyly
- Hypospadius
- Cleft palate
- Heart murmur
- Cyanosis and/or respiratory distress secondary to cardiac and pulmonary anomalies (4 most common lesions: atrial septal arteriosus, endocardial cushion defects, anomalous pulmonary venous return and patent ductus arteriosus$^2$)
was initially reluctant to use this device. However, upon further discussion, a consensus was reached that this was the best alternative available for the patient at this time. The LMA was placed using the standard index finger insertion technique. The patient immediately responded to ventilation through the LMA as demonstrated with observation of good chest excursion, auscultation of adequate breath sounds, and improved ABGs (see Table 2).

With an airway device secure and functioning well, the patient was transported to a tertiary facility where he was evaluated by an otolaryngologist and received an emergency tracheotomy. A full evaluation of the patient concluded with the diagnosis of SLOS type II. The patient initially thrived and plans were made for his discharge. Unfortunately the infant died the evening before going home at age 36 days.

Discussion
Patients diagnosed with SLOS have the potential to be difficult to intubate. In a series of 3 patients reported by Choi and Nowaczyk,1 1 patient had an abnormal laryngoscopic view, which worsened with age. Haji-Michael et al reported 1 difficult intubation in his series of 5 patients.7 Quezado et al, in a series of 20 anesthetics on 14 patients, had 5 patients with a history of difficult intubation including 1 procedure that was aborted due to failure to intubate.8 Concern should exist for difficult intubation with any patient diagnosed with SLOS. Prominent incisors, micrognathia (67%-100%), tongue abnormality (12%-63%), and palate abnormalities (78%) are common.1 These anatomic features increase the difficulty of laryngoscopy and intubation.

Airway management during neonatal resuscitation following the American Heart Association guidelines is achieved by bag mask ventilation or intubation. Occasionally bag mask ventilation can be difficult and tracheal intubation impossible, as was the case with the patient in this case report. Several studies have been published regarding the use of the LMA in children; however, the experience in using the LMA in infants is limited and scarce in the premature neonate. Cadaveric studies in infants demonstrate that, despite anatomic differences between adults and infants, the current design of the LMA Classic is appropriate for infants.9 Size 1 is recommended for use in infants weighing 2.5 to 5.0 kg. Two case series by Paterson et al10 and Gandini and Brimacombe11 have been published to support the safe and effective use of the LMA in neonatal resuscitation. The LMA provides a clinically patent airway for positive pressure ventilation, continuous positive airway pressure, and spontaneous breathing.12

The possibility for injury exists with any invasive airway management technique, including intubation and LMA insertion. Esmail et al studied a group of 40 neonates and reports that epiglottic trauma was present in 2 cases with LMA and in 1 case with endotracheal tube. Trauma to the uvula occurred in 4 cases with the LMA and in 2 cases with the endotracheal tube.13 However, when repeated intubation attempts by one or more providers fail, surely the risk of airway trauma will increase. Introduction of the LMA early in the airway management algorithm minimizes any further injury to already violated tissues. Referring to Poiseuille’s law for laminar flow and the critical relationship between tube radius and flow resistance \( R=\frac{8nL}{\pi r^4} \),14 edema that might reduce the radius of the airway by half would increase airway resistance 16 times! The already small airway of the neonate, and even smaller in the premature neonate, does not allow much room for edema. The neonate in fragile condition, requiring resuscitation, may not tolerate the increases in airway resistance. Early placement of the LMA, after unsuccessful intubation attempts, has the potential to improve respiratory care.

Insertion of the LMA is easy to learn. Nurses, residents, midwives, and emergency medical technicians have all been successfully trained to place LMAs in resuscitative efforts within 30 seconds.15 Morse et al concluded that a short training period (a demonstration and practice 5 times) enabled dental students to rapidly and effectively place the LMA.16 Coulson et al compares the LMA insertion times with inexperienced personnel and reports a first attempt success rate of 85%.17

In the present case report, the pediatrician only had experience placing the LMA on a manikin, yet with verbal coaching was able to successfully place the LMA. In the rural setting, where there is limited or no anesthesia service, the skill of LMA placement by nonanesthesia professionals may very well save lives, including those within the neonatal population. Transport to a tertiary facility is often delayed until an adequate airway is established. In this case report, the

### Table 2. Arterial blood gas results

<table>
<thead>
<tr>
<th>Time</th>
<th>pH</th>
<th>PCO₂</th>
<th>PO₂</th>
<th>HCO₃⁻</th>
</tr>
</thead>
<tbody>
<tr>
<td>Before laryngeal mask airway (LMA) placement</td>
<td>12:37 PM</td>
<td>7.14</td>
<td>56</td>
<td>128</td>
</tr>
<tr>
<td></td>
<td>1:48 PM</td>
<td>7.18</td>
<td>63</td>
<td>111</td>
</tr>
<tr>
<td>After LMA placement</td>
<td>2:35 PM</td>
<td>7.32</td>
<td>43</td>
<td>161</td>
</tr>
<tr>
<td></td>
<td>3:00 PM</td>
<td>7.35</td>
<td>40</td>
<td>251</td>
</tr>
</tbody>
</table>
LMA Classic size 1 contributed to the successful stabilization and transport of this patient.

REFERENCES


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Yolanda R. Leal-Pavey, CRNA, BS, is staff nurse anesthetist at Deaconess Medical Center in Spokane, Wash. The event reported in this article occurred when the author was an independent practitioner in rural Colorado.