A rare occurrence of pyloric stenosis in an infant with osteogenesis imperfecta: Anesthetic implications

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Abstract
Congenital anomalies pose many challenges during anesthesia due to anatomic and physiological alterations. The inherent complications associated with the disorders necessitate vigilance for providing anesthesia to even seemingly simple surgical intervention. Here, we share our experience of anesthesia management of an infant of congenital osteogenesis imperfecta with pyloric stenosis for pyloromyotomy.

Key words: Anesthesia, infant, osteogenesis imperfecta, pyloric stenosis, pyloromyotomy

Introduction
Infantile hypertrophic pyloric stenosis (IHPS) was first described by Hirschsprung in 1888. The prevalence of IHPS ranges from 1.4 to 8.8 cases/1000 live births. The condition usually presents at 1-2 months of age. The classical presenting features are non-bilious, projectile vomiting, visible peristalsis and hypochloremic hypokalemic metabolic alkalosis. Schwartz et al. have reported in a retrospective study that 36% of babies presented with hyperkalemia. 6-10% of babies may have associated congenital anomalies such as cleft palate and esophageal reflux.

Osteogenesis imperfecta (OI) is a rare congenital anomaly occurring in 1:50,000 births. It is a connective tissue disorder caused by a defect in collagen Type 1, affecting bone, teeth, sclera and ligaments. The Sillence classification describes four types. Type 1 is a mild form of OI, Type 2 is most severe and occurring perinatally, Type 3 is progressively deforming and Type 4 has mild to moderate bone fragility. The coexistence of these two conditions is very rare with scarcity of literature. We report a case of 2-month-old infant of OI with pyloric stenosis for pyloromyotomy.

Case Report
A 2-month-old infant presented with non-bilious projectile vomiting following breast feeds since 15 days and constipation since 8 days. The baby was lethargic, moderately dehydrated with sunken eyeballs, had poor skin turgor and depressed anterior fontanelle. His birth weight was 3 kg, which had presently reduced to 2.5 kg.

He was pale with a characteristic blue sclera [Figure 1]. There was bowing of all four extremities, but no facial or chest abnormalities [Figure 2]. His abdomen showed visible peristalsis and a firm mass palpable in the epigastrium. The cardiovascular and respiratory system was unremarkable.

His infantogram showed a multiple healed long bone fractures in both extremities [Figure 3]. Ultrasound abdomen showed hypertrophy of the pyloric musculature causing stenosis.

Based on above findings, he was diagnosed as pyloric stenosis with OI with moderate dehydration.

Investigations revealed hemoglobin of 11.7 g/dL, platelets 383 K/uL, serum creatinine 0.7 mg/dL, blood urea nitrogen 10.9 mg/dL, electrolytes were Na+ 125 mmol/L, K+ 2.8 mmol/L and Cl− 74 mmol/L. Thyroid function tests were within the normal limits. His arterial blood gas revealed a pH of 7.619, pCO₂ of 38.5 mm Hg and HCO₃⁻ of 38.5 mmol/L.
Medical management was done in the form of correction of dehydration and electrolyte imbalance over a period of 48 h using 5% dextrose in 0.45% saline and potassium chloride. His pre-operative electrolytes were Na⁺ 133 mmol/L, K⁺ 4.2 mmol/L and Cl⁻ 91 mmol/L and pre-operative blood sugar was 100 mg%. Operation theater was prepared anticipating difficult airway, with Miller and Macintosh laryngoscope blades, endotracheal tubes, stylet and laryngeal mask airway (LMA). Electrocardiogram and SpO₂ monitors were attached. Ryle’s tube aspiration was performed. The baby was preoxygenated for 5 min and premedicated with intravenous (IV) glycopyrolate. General anesthesia was induced with thiopentone sodium 5 mg/kg and atracurium 0.5 mg/kg. Trachea was intubated with a modified rapid sequence induction technique using a 3.5 mm endotracheal tube with cricoid pressure and gentle laryngoscopy. Cormack Lehane grade of laryngoscopic view was II. Capnograph and temperature probe were attached. An additional IV access was secured for replacement fluids. Care was taken to avoid pressure sores and fractures by applying cotton pads to pressure points. Intraoperatively anesthesia was maintained with oxygen, air and sevoflurane (1-1.5%). Local infiltration with 0.1% bupivacaine and paracetamol suppository was used for post-operative analgesia.

A pyloromyotomy was done using a right upper quadrant transverse incision. He received 25 ml of 1% dextrose in ringer lactate over the duration of the surgery, which lasted 45 min. His heart rate was maintained between 120 and 140/min; SpO₂ 97-99% and temperature between 35.5°C and 36.5°C during this period.

The neuromuscular blockade was reversed with glycopyrolate 8 mcg/kg and neostigmine 0.05 mg/kg. Post-extubation, he had a good cry and was shifted to the pediatric intensive care unit (PICU) for post-operative monitoring and fluid replacement. His post-operative electrolytes showed Na⁺ 129 mmol/L, K⁺ 5.0 mmol/L and Cl⁻ 98 mmol/L and appropriate electrolyte correction was done with maintenance of input output charting.

He started taking oral feeds on the 2nd post-operative day. He was shifted out of the PICU on the 5th day and discharged on the 7th post-operative day without any complication.

**Discussion**

Pyloromyotomy is the definitive and curative treatment for pyloric stenosis. Before surgery, it is important that the infant is adequately hydrated, with correction of electrolyte abnormalities and metabolic alkalosis. If not corrected, compensatory respiratory acidosis coupled with residual anesthesia may affect recovery from general anesthesia and increase the risk of post-operative apnea.

Our patient had moderate dehydration with hypokalemic, hypochloremic metabolic alkalosis, which was corrected with 5% dextrose in 0.45% saline and potassium replacement. Patients
of OI have a greater incidence of airway anomalies, thoracic wall abnormalities, platelet dysfunction, hyperthyroidism and increased tendency for perioperative hyperthermia. They may have megalocyphaly, macroglossia, cranio-vertebral junction problems including atlanto-axial subluxation and short neck contributing to a difficult airway.\[4,5\]

These patients must be handled very gently. Fractures may occur due to simple procedures such as taking blood pressure, applying tourniquet and positioning on the operation theatre table. Airway management may also cause fractures.

Occasionally, visualization of the airway is difficult and use of LMA may be very useful.\[6\] Infants for pyloromyotomy are at greater risk of aspirating gastric contents, this demands a secured airway with endotracheal intubation. Pre-operatively thorough evacuation of stomach contents through a nasogastric or orogastric tube should be done. Cook-Sather et al. compared three techniques: Awake intubation, rapid sequence intubation and modified rapid sequence intubation. Awake intubation neither prevented bradycardia nor oxygen desaturation and is no longer used. As these babies are usually low weight they do not tolerate complete apnea even for 60 s during the rapid sequence intubation.\[7\]

We used modified rapid sequence technique, which consisted of pre-oxygenation followed by IV induction and non-depolarizing muscle relaxant. Gentle positive pressure ventilation was provided along with cricoid pressure prior to endotracheal intubation. Air, oxygen and sevoflurane were used for maintenance. In these infants of OI succinylcholine is avoided, as succinylcholine induced fasciculation can cause fractures and dislocation. They also have a higher propensity for malignant hyperthermia. Intraoperative hyperthermia has been reported, which may be due to associated hyperthyroidism and increased metabolic rate. The routine pediatric anesthesia practice of preventing intraoperative hyperthermia should be used judiciously with continuous temperature monitoring.

Maintenance of anesthesia usually consists of inhalational agents. Desflurane has an advantage because of its rapid elimination and is useful in these infants who are prone to apnea and ventilatory depression.\[8\] Opioids should be avoided. If at all needed, remifentanil is the drug of choice because of its useful characteristics.\[9\] Local infiltration with bupivacaine and rectal paracetamol provides adequate analgesia in most patients. Infant should be fully awake with intact protective reflexes, and have a regular breathing pattern before extubation. Adequate splinting of arms and legs must be ensured to minimize postoperative pain and avoid excessive limb movements for prevention of any additional fractures.

Incidence of post-operative apnea is higher; hence apnea monitoring along with pulse oximetry is indicated for first 24 h after surgery, irrespective of the drug used.\[10\] Parenteral fluids are administered to maintain adequate hydration until oral intake is sufficient.

Thus to conclude, we recommend extra gentle care while handling these babies to prevent the complications such as fractures of bones, atlantoaxial dislocation, excessive bleeding and hyperthermia.

Pyloric stenosis is a medical emergency and should not be converted into a surgical nightmare by premature surgical repair before adequate resuscitation.

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How to cite this article: Jagtap SR, Bakhshi RG, Jain A. A rare occurrence of pyloric stenosis in an infant with osteogenesis imperfecta: Anesthetic implications. J Anaesthesiol Clin Pharmacol 2014;30:270-2.

Source of Support: Nil, Conflict of Interest: None declared.