Profound Hypoxemia during Major Abdominal Surgery in a Small Infant with Tetralogy of Fallot

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We report a protracted hypoxic event during Soave's endorectal pull-through for Hirschsprung's disease in a 2-month-old male infant with tetralogy of Fallot. After the bowel loops were delivered out of the abdominal cavity, profound hypoxemia occurred which persisted for about 120 min. The hypoxemia was completely resolved after the intestine was reduced back into the peritoneal cavity. The immediate cause was postulated to have been decreased systemic vascular resistance. Associated factors included hypothermia and acidosis leading to high pulmonary vascular resistance which further aggravated the right-to-left blood-shunting situation in this patient. Postoperative follow-up showed no neurological complications. Small infants with complex heart disease should be carefully evaluated before major abdominal surgery. *(Chang Gung Med J 2005;28:498-502)*

Key words: tetralogy of Fallot, profound hypoxemia, major abdominal surgery.

Tetralogy of Fallot (TOF), the most-common cyanotic congenital heart disease, consists of a ventricular septal defect, right ventricular outflow tract obstruction, an overriding aorta, and right ventricular hypertrophy. Tetralogy of Fallot presents in various patterns from a pink TOF to a very complicated one.(1) The severity of pulmonary outflow tract obstruction, which increases with tachycardia and an inotropic state, and decreased systemic vascular resistance (SVR), plays a major role in the degree of shunting. Transient hypercyanotic spells are not rare during anesthesia in patients with TOF and usually are reversible without serious complications.(2) However, prolonged life-threatening hypoxemia during major abdominal surgery or a warning of such a crisis has not been presented in the medical literature. We report an unusual experience of a protracted hypoxic event during major abdominal surgery in a 2-month-old infant with TOF. The possible mechanisms and its management are discussed.

CASE REPORT

A male baby, born at 39 weeks of gestational age with a birth body weight of 3050 g, was admitted at 4 days old for progressive abdominal distention and vomiting. An emergent loop ileostomy and multiple biopsies of the rectum and colon proved a diagnosis of long-segment Hirschsprung's disease. The anesthesia course was smooth with no record of hypoxemia. This infant was found to have TOF by 2-dimensional echocardiography soon after birth. The pulmonary artery was hypoplastic. The postoperative course was smooth with no sequelae. Elective Soave's endorectal pull-through procedure was scheduled for Hirschsprung's disease at 2 months of age.

According to his parents' description, this male

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infant had mild cyanosis with irritability and crying, but no definite blue spell episode was recognized. At the preanesthetic examination, he looked weak but not cyanotic. There was no need for oxygen therapy or ß-blockers. Preoperative laboratory findings were within normal limits; hemoglobin was 12.1 gm/dl and hematocrit was 33.6%. The chest X-ray showed increased perihilar infiltration on both sides, widening of the upper mediastinum, and mild cardiomegaly.

In the operating room, the first recorded peripheral oxygen saturation (SpO_2) of this baby breathing spontaneously with room air was 86%. The baseline blood pressure was 64/36 mmHg and heart rate was 144 beats/min. General anesthesia was induced using sevoflurane with oxygen by facemask. Rocuronium bromide (0.6 mg/kg) was used to facilitate the tracheal intubation. After successful induction, a central venous line was set up via the right internal jugular vein and an indwelling intra-arterial catheter was established at the left radial artery with a 24-gauge catheter for continuous monitoring of the hemodynamic status and blood sampling. Anesthesia was maintained with sevoflurane as the main anesthetic regimen and rocuronium bromide as the paralytic agent.

The patient's condition remained stable until the surgeon pulled out all of the intestinal loops, at which point the oxygen saturation shown on pulse oximetry (SpO_2) abruptly decreased, and the arterial blood pressure also gradually decreased. Under the suspicion of inadequate circulating blood volume, increased-speed fluid resuscitation and a dopamine drip were given. In spite of these interventions, the patient's condition did not improve. Further deterioration of the SpO_2 was noted and dropped to only 23%. The intestine showed poor perfusion and the heart rate rose to 180 beats/min. Intravenous esmolol (with a loading dose of 500 µg/kg and then a 50 µg/kg/min continuous infusion) was begun. Another 4 bolus doses of esmolol were given to control the tachycardia and to decrease the infundibular spasms. The inhalation anesthetic was discontinued and a large dose of morphine sulfate was given to blunt the hemodynamic response and to decrease the pulmonary vascular resistance (PVR). Blood gas analytical data showed metabolic acidosis, which was corrected by sodium bicarbonate. Concomitant hypothermia (35.5°C at the nasopharynx) was detected, and an overhead infrared radiation warmer was applied. With all these efforts the SpO_2 rose to 50%~60%, the systolic blood pressure was around 60 mmHg, and the heart rate was maintained below 140 beats/min. This hypoxic condition had now persisted for about 120 min.

As soon as the intestine was reduced back into the abdominal cavity and the fascia was being closed, the SpO_2 climbed up to 95% and remained there throughout the rest of the surgery. No further desaturation episodes took place. The vital sign parameters were stable. The operation lasted for 180 min, and the baby was sent to the pediatric intensive care unit for further care.

Postoperatively, the vital signs were quite stable and the oxygen saturation was kept at 80%~85% under a fraction of inspiratory oxygen (FiO_2) of 30%, and was able to achieve 85%~90% with an FiO_2 of 100%. Intermittent desaturation episodes were found and were improved by positive ventilation using an Ambu bag with an increased oxygen flow rate. He was gradually weaned off ventilator support, and esmolol infusions after approximately 24 and 48 h, respectively. The infusion rate of the esmolol was gradually tapered down and shifted to oral propranolol when he could tolerate oral feeding. He was successfully extubated 24 h after the end of surgery and made a full recovery with no sequelae. He tolerated regular oral feeding 4 days after surgery and was transferred to the ward for regular postoperative care 7 days after the event. He was discharged home on the tenth postoperative day. He was kept on oral propranolol treatment at home.

For the postoperative neurological evaluation, a brain sonographic examination was done via the anterior fontanel on the sixth day after surgery, and data showed no definite pathology except for some brain swelling. A subsequent examination of the baby disclosed a good response to stimulus for his age, and he was able to actively move about. No neurological damage was found. At a follow-up 2 weeks after the hypoxic crisis episode, this baby was in an acceptable condition with no neurological sequelae.

DISCUSSION

Although the clinical safety of anesthesia has dramatically improved in recent years, small infants with complex cyanotic heart disease undergoing

major non-cardiac surgery are still a challenge for most anesthesiologists. Morray et al. from the Pediatric Perioperative Cardiac Arrest Registry report showed that patients younger than 1-year-old with congenital heart disease have the highest predicted rate of cardiac arrest during surgery.[3]

The basic pathophysiology of TOF is related to the degree of right ventricular outflow tract obstruction.[4] In the anesthetic management of patients with TOF, a preoperative assessment should pay particular attention to any obstruction of the right ventricular outlet. The importance of a careful review of the cardiac catheterization data and complete understanding of its potential impact on the operative and anesthetic plan are essential. The main anesthetic consideration is to maintain the PVR/SVR ratio within a normal range to prevent aggravation of right-to-left shunting. Any situation that might decrease the SVR should immediately be corrected. Intraoperative hemodynamic goals include stabilizing the heart rate, increasing the contractility, increasing the preload, decreasing the PVR, and maintaining a normal SVR to achieve optimal perfusion and oxygenation.[2] Successful resuscitation by compression of the abdominal aorta from near-fetal hypercyanotic episodes has been reported.[5,6] During open-chest operations, surgeons may briefly clamp the thoracic aorta to reverse hypoxic spells.[7]

In our case, it is reasonable to assume that the hypoxic event was likely attributable to the sudden decrease in the SVR after the intestinal loops were pulled out of the abdominal cavity. This supposition was supported by the dramatic improvement in the oxygen saturation after the intestine was reduced back into the abdomen. The increased heart rate during the surgery induced poor right heart filling and further deteriorated the cardiac output. Hypoxemia, hypothermia, and metabolic acidosis were all contributing factors for the increased PVR and the exacerbating ventricular infundibular spasms. In brief, 5 mechanisms might have affected the arterial hypoxemia in this patient: (1) decreased SVR, (2) increased PVR, (3) right ventricular infundibular spasms, (4) increased right-to-left shunting, and (5) hypovolemia.

A potent inhalation agent was chosen for this patient as the principal anesthetic agent for it allowed early postoperative extubation and avoided a prolonged period of intensive care. After the hypoxic event, the anesthetic agent was shifted to morphine because morphine lowers the PVR. Most inhalation agents have a vasodilating effect and will decrease the SVR. The high-dose opioid technique remains popular for pediatric cardiac surgery. Respiratory depression and excessive sedation in the postoperative period may be a problem if opioids are given in high doses to infants, but that argument is not applicable if the newer opioids such as alfentanil and remifentanil are used, because of their much-shorter half-lives.[8,9]

Based on the above findings, the profound hypoxic event might have been avoided by a prophylactic increase in the SVR such as with a restrictive bandage on the lower limbs, movement to Trendelenburg's position, appropriate intraperitoneal compression to maintain appropriate abdominal pressure, and use of a rational pharmacological approach to optimize the heart rate.[10] On the other hand, palliative procedures for TOF, such as systemic-to-pulmonary arterial shunts, percutaneous pulmonary valvuloplasty,[11] or even complete corrective surgery may be considered before major bowel surgery.

Fortunately, the patient developed no neurological impairments clinically. A prolonged severe hypercyanotic crisis can provoke catastrophic outcomes secondary to tissue hypoxia such as encephalopathy, myocardial dysfunction, and renal damage.[12] Patients with cyanotic congenital heart disease may possibly have a higher tolerance for hypoxic stress and this may explain the better outcome in this young infant.[13]

In conclusion, the hard-learned experience in this case stresses the possibility of serious consequences during major abdominal surgery in a small infant with complex congenial heart disease. In current medical care, increasing numbers of major non-cardiac surgeries are being performed on small infants with high anesthetic risks such as extreme prematurity, complex heart disease, or other congenital anomalies. The attending anesthesiologist should have good strategies to cope with these challenges. The decision-making for surgery in high-risk infants should be individualized, and anesthetic management must depend on the patient's clinical presentation and surgical variables.

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一法洛氏四重症嬰兒於大腹部手術中發生嚴重血氧飽和度下降

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報告一兩個月大、患有法洛氏四重症先天性心臟病之嬰兒，因巨大結腸症進行 pull through 手術中發生嚴重血氧飽和度下降情形。手術進行至腹腔內臟往外翻出後病人發生嚴重血氧飽和度下降、持續約 120 分鐘之久。缺氧情形於內臟放回腹腔後完全獲得改善。發生原因推想可能爲內臟往外翻出後腹壓降低，體循環阻力突然減低，加上體溫低、酸血症等因素，使從右至左血液分流變得很厲害，因而發生嚴重血氧飽和度下降情形。術後病人沒有明顯神經系統方面受傷情形。建議有嚴重先天發紺性心臟病病人，進行大型侵犯性之腹部手術術前應予以慎重評估。(長庚醫誌 2005;28:498-502)

關鍵字：法洛氏四重症，嚴重缺氧，腹部手術。

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