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Anesthetic Management for Concomitant Correction of Congenital Cardiac Defects and Long "O" Ring Tracheal Stenosis: A Role for Heliox?: Report of 2 Cases

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We present 2 infants with the rare association of long congenital tracheal stenosis, ventricular septal defect, and pulmonary hypertension. We describe a step-by-step assessment of the patients and the necessary procedures for a successful concomitant repair of both cardiac and tracheal malformations. The use of a helium–oxygen mixture (heliox) for the induction of anesthesia and pre-cardiopulmonary bypass is discussed. (A&A Practice. 2018;10:154–6.)

Congenital heart disease correction is often a challenge in children. However, with an anatomic correction, most patients can hope to lead a normal life.[1] The same applies to congenital tracheal stenosis (CTS) with complete "O" tracheal rings. CTS was classically described by Cantrell and Guild[2] and was classified into 3 types (Figure 1): type 1 is generalized hypoplasia (67% mortality rate), type 2 is funnel-type stenosis (29% mortality rate), and type 3 is segmental stenosis with 2 or 3 cartilage rings involved (no mortality).[3] Advances in surgery have allowed these children to survive and grow normally. The tracheal airway enlarges in age-appropriate dimensions with the growth of the child.[4] The "O" ring tracheal segment is corrected by a slide tracheoplasty (ST), wherein the narrow tracheal segment is transected in the middle; the 2 cut ends are split in the front and back and then are slid over each other. Thus, the length of the stenotic segment reduces by half, but the inner diameter doubles and the resistance to air flow decreases dramatically. For this procedure, cardiopulmonary bypass (CPB) is mandatory. Any tracheal instrumentation causing edema could transform an already compromised airway into a critical airway, and hypoxia or hypercarbia could cause right ventricular failure. We present 2 children with ventricular septal defects (VSDs) and pulmonary hypertension combined with a severe tracheal stenosis in which the administration of a helium–oxygen mixture (heliox) increased the margin of safety.

Written consent from the legal guardian was obtained for both patients.

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CASE DESCRIPTIONS

Case 1

A 2-year-old boy was planned for surgical correction of a large VSD. On examination before surgery, he presented with significant stridor. The chest computerized tomography scan revealed a CTS with a diameter of 3 mm. Rigid endoscopy performed by Ear, Nose, and Throat team confirmed the diagnosis of type 1 stenosis (Figure 1) with complete rings extending from the first tracheal ring up to the carina (Supplemental Digital Content, Video, http://links.lww.com/AACR/A154). The endoscopic evaluation was done on a spontaneously breathing patient after sevoflurane induction and propofol maintenance without any complication. Transthoracic echocardiography revealed a large unrestricted VSD with bidirectional shunt having a low gradient, suggesting elevated pulmonary resistances. To assess the reversibility of this pulmonary hypertension, the child was referred for cardiac catheterization. He was induced with sevoflurane, and a laryngeal mask airway was inserted for the duration of the anesthetic. On spontaneous-assisted ventilation, obtaining tidal volumes of 5–8 mL/kg required high ventilation settings (positive pressure of 35–40 cm H_2O). Because of these, we considered heliox for subsequent anesthetics. Despite a $Paco_2$ above 50 mm Hg, the child remained hemodynamically stable. Oxygenation was never an issue with peripheral oxygen saturation (Spo_2) above 95% at room air. The systolic pulmonary pressure was isosystemic on inspiratory oxygen fraction (Fio_2) 0.21, with good reversibility on Fio_2 1.0 and 20 ppm inhaled nitric oxide.

VIDEO+

Case 2

The second boy was 16 months old and presented with the same symptomatology. The tracheal stenosis with complete rings had an inner diameter of 2 mm and extended from the first tracheal ring up to the carina. He also had a large VSD, an atrial septal defect, but only slightly elevated pulmonary pressures. Hence, the cardiac catheterization was not performed.

Cases 1 and 2

Both patients were planned for sequential cardiac and tracheal surgeries under CPB. They were induced with sevoflurane using a Infant Mapelson F circuit (Jackson-Rees, Covidien, Mansfield) We added side tubing connected to a 78% helium–22% oxygen tank provided by industry to

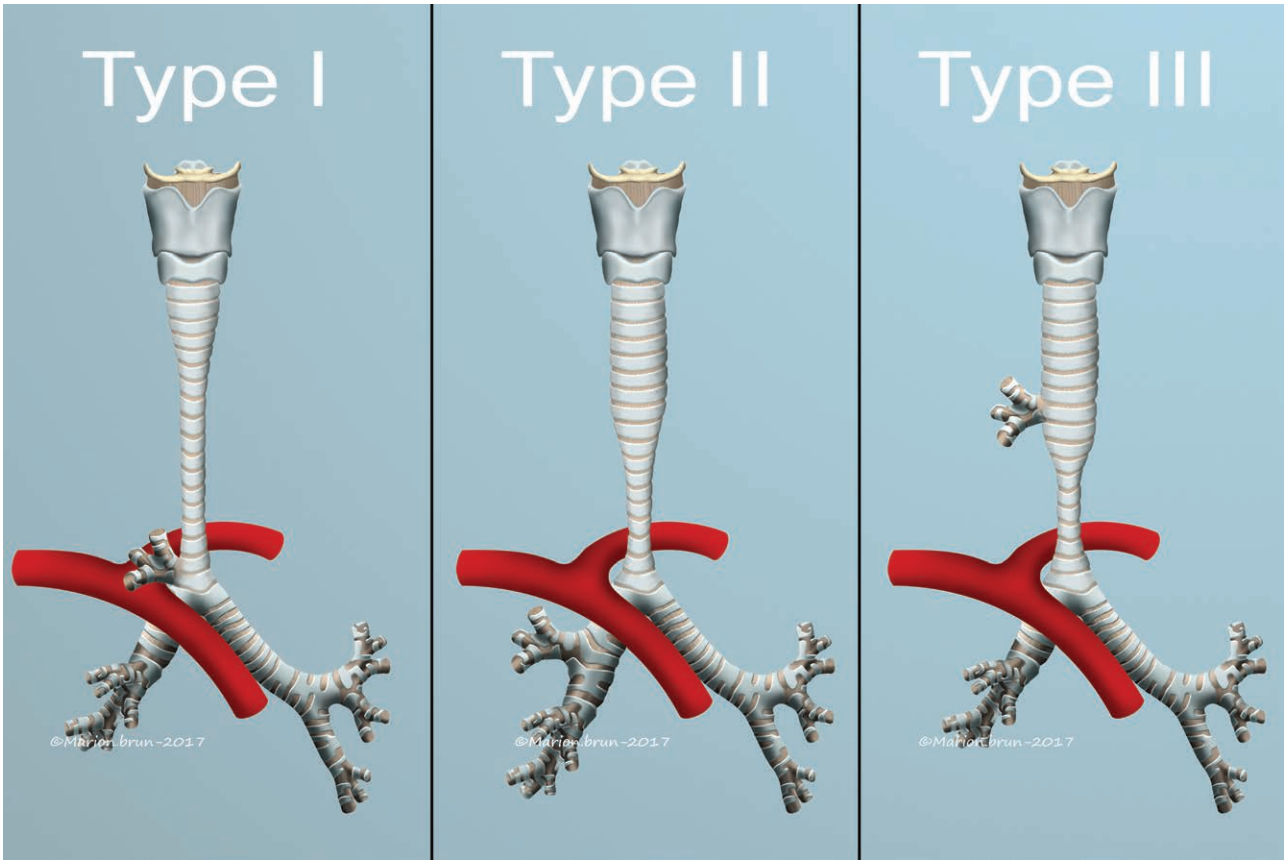

Figure 1. Classification of congenital tracheal stenosis with circular "O" rings according to Cantrell and Guild.[2]

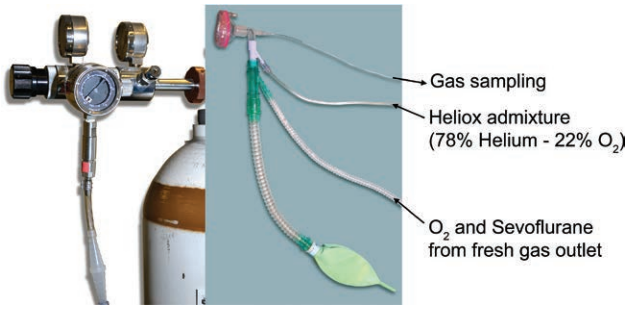

Figure 2. Jackson-Rees combined with heliox setup. O_2 indicates oxygen.

decrease density of the inhaled gases (Figure 2) and used this gas mixture from induction until CPB was fully established.

After central venous and arterial lines were placed, propofol was started for maintenance of anesthesia and morphine was carefully titrated. At the same time, the cardiovascular team performed a femoral cannulation to initiate temporary CPB, a rescue plan in case of airway loss caused by deep anesthesia and paralysis for sternotomy. Despite adequate cannulae placement, reduced inflow limited CPB to half of theoretical full support (based on cardiac index of 2.5 L/min/m^2)[5] for these children. We managed to insert an endotracheal tube (ETT) (Portex; Smith Medical, 4.0 mm in the first child; and a 3.5-mm Microcuff; Halyard Health, in the second child) just below the vocal cords and above the stenotic trachea. The cuffed tube in the second child was a better option and resulted in smaller leaks, better positive end-expiratory pressure, and more accurate measurement of $Etco_2$. Both patients remained hemodynamically stable with Spo_2 98%–100% on manually controlled ventilation (Fio_2, 0.4; peak positive pressure, 25–30 cm H_2O; positive end-expiratory pressure, 5 cm H_2O; respiratory rate, 20/min; tidal volume, 8 mL/kg).

After CPB was fully established with central cannulations, the surgical teams performed cardiac and tracheal repair. With an enlarged tracheal diameter after the ST, uncuffed ETTs were intraoperatively passed more distally up to the mid-trachea, and the tube position was confirmed endoscopically (3.1-mm Olympus Video Bronchoscope BF-XP190 through the ETT; Tokyo, Japan) in relation to the carina. The intraoperative course was uneventful. At the end of the surgery, the children were transferred intubated to the intensive care unit and extubated on the fourth and fifth day, respectively.

DISCUSSION

Long-segment CTS is a rare malformative disorder typically presenting in the first years of life with acute respiratory failure. The anesthetic management aims to ensure proper oxygenation and ventilation, secure the airway, and allow diagnostic or therapeutic procedures. However, any mucosal damage or manipulation during endoscopy can lead to severe airway edema and acute obstruction. In this setting, we favor assisted spontaneous ventilation with sevoflurane induction and minimal airway instrumentation. Mechanical ventilation is much more efficient, however, with the positive pressure

helping to maintain the patency of the distal malacic tracheobronchial tree, eliminating the risk of gastric insufflation, and allowing a better control of $Paco_2$. But atraumatic intubation is impossible in the context of these extremely narrow tracheas.

In our cases, airway control was achieved by passing an ETT through the vocal cords and up to the stenotic trachea. It allowed us proper oxygenation and ventilation, and we did not have to face any alarming desaturation. As a backup plan, we were prepared for a tracheal intubation with a very thin 8F Cook Airway Exchange Catheter (Bloomington, IN) for oxygenation that would have been passed directly through the stenosis. Fortunately, this was not necessary.

Several types of tracheal reconstructions have been proposed since Kimura et al[6] in 1982, with ST appearing to be preferable to a pericardium patch or cartilage tracheoplasty.[7] Associated malformations are very common and can be found in up to 84% of patients,[8] with cardiac or vascular malformations seen in 70%.[9] Our patients presented with pulmonary hypertension, and this feature increases strongly the anesthetic risk.[10] Any elevation in $Paco_2$ or hypoxemia can lead to acute right ventricle failure. We took the option not to instrument the airway during bronchoscopy and catheterization to protect the mucosa, but at the expense of high driving pressures. In this context, we decided to add helium to reduce inhaled gas density. This has been described by several authors since Barach[11] in 1934. Recently, a very interesting case of airway obstruction in an adult patient was reported by Galway et al.[12] Their management was very similar to ours, because they favored spontaneous ventilation until the airway was secured and added heliox to decrease work of breathing.

Helium has a density nearly 6 times lower than air. Based on a computational fluid dynamics model showing a reduction of pressure drop across a constriction to one-third of its value with air,[13] we hypothesized that the addition of helium in our patients would help reduce the peak pressures during assisted ventilation. We used a basic Jackson-Rees circuit powered by the anesthetic machine for oxygen and volatile anesthetics delivery with a side tubing providing a 78/22 helium/oxygen mixture from an accessory tank. Measurement of inspired helium fraction being technically very complicated, we assumed that it was 1 minus the FIO_2 measured by an in-line oxygen analyzer whose main purpose was to avoid administration of an hypoxic mixture because of stratification of the highly compressed gases (as heliox). This configuration also allowed control of the tidal volume by direct hand ventilation and chest expansion observation. Although semiclosed circuits for helium administration would be much more cost-effective according to Jurickova et al,[14] the relative short-term use and the risk of volume-induced injury made us favor our custom-made open circuit. Furthermore, not every ventilator is Food and Drug Administration approved for delivery of heliox.[15]

In conclusion, a helium and oxygen mixture can be considered as a useful addition to the anesthesia strategy while concomitantly managing a congenital cardiac and long-segment "O" ring tracheal stenosis. In computational models, helium reduces the pressure drop across an airway constriction, and thus should help reduce the pressure needed for assisted ventilation. Protection of the tracheal mucosa is essential, and spontaneous ventilation with minimal airway instrumentation is the best, at the expense of high driving pressures. However, when required, insertion of an ETT just above the stenosis is feasible and may be lifesaving.

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