Anaesthetic management of severe scoliosis correction in spinal muscular atrophy, severe restrictive lung disease and difficult airway

Sir,

Scoliosis correction can be associated with severe intraoperative blood loss and a high rate of anaesthetic and surgical complications. We report a case of spinal muscular atrophy type II (SMA II) posted for scoliosis correction. The patient had severe restrictive lung disease and a difficult airway that caused significant challenges to the anaesthesiologist.

A 32-year-old woman, weighing 35 kg and who was a known case of SMA II, was electively posted for scoliosis correction surgery. Her Cobb’s angle was 53° and she had associated paraparesis. The goal of the surgery was to improve the sitting balance of the patient on a wheelchair. Her thoracic cavity was protruding anteriorly. Airway examination revealed inter-incisor distance 3 cm, Mallampati class IV and severely restricted neck movements. Pulmonology evaluation showed severe restrictive lung disease with forced expiratory volume in the first second (FEV₁) 37% of predicted. Preoperative echocardiogram and blood investigations were normal. Preoperative arterial blood gas (ABG) analysis showed hypoxia with partial pressure of oxygen (PaO₂) of 81 mm Hg and normocapnia [Figure 1].

In the operation theatre, with standard monitoring, she was intubated using C-MAC videolaryngoscope without giving muscle relaxant, although we had prepared well for a difficult airway including awake fibreoptic intubation. Tranexamic acid infusion at 150 mg/h was given during surgery after 500 mg bolus as suggested by the surgeon. Muscle relaxant was given before turning the patient prone and traction was applied. Thereafter, the peak inspiratory pressure (P peak) increased to 40 cm H₂O and tidal volume (Vₜ) decreased to 180 ml. ABG analysis revealed severe respiratory acidosis with pH 6.9 and 7.3, partial pressure of carbon dioxide (PaCO₂) 115 mm Hg and 56 mm Hg at the first hour and third hour of surgery respectively, and normal PaO₂ values. All the mechanical causes of hypercarbia were ruled out. High minute ventilation was kept to reduce end-tidal carbon dioxide (EtCO₂). Since respiratory acidosis persisted, it was decided to stage the surgery. Vₜ and P peak became normal soon after the patient was made supine, which might have been due to improvement in lung compliance. Extubation was done on the following day after correction of acidosis and hypothermia.
On the second postoperative day, the patient complained of loss of vision in the right eye and this was diagnosed as central retinal artery occlusion by the ophthalmologist. The use of tranexamic acid, increased pressure on eye globe and skull traction were thought to be the risk factors for postoperative vision loss (POVL).

During the second stage surgery, skull traction was avoided (after discussing with the surgeon). Moreover, gel head-ring was used instead of foam headrest, tranexamic acid was avoided, soft bolsters were used instead of firm ones to improve lung compliance, and the duration of surgery was reduced. Intermittent eye massage was given to the unaffected eye to improve blood circulation as suggested by the ophthalmologist. The second stage surgery was completed with minimal hypercarbia and no further complications. Postoperatively, the patient was able to attain sitting balance on wheelchair and her pulmonary function tests also improved [Figure 2].

In this case, we encountered hypercarbia and an increased $P(a-Et)$CO$_2$ difference. The increase in $P(a-Et)$CO$_2$ could have been due to increased ventilation-perfusion mismatch (V/Q mismatch), positive pressure ventilation, especially with positive end-expiratory pressure (PEEP), and high respiratory rate-low $V_T$ ventilation. If the CO$_2$ elimination is constant, PaCO$_2$ is inversely related to alveolar ventilation. Inefficient ventilation indicates increased dead space to $V_T$ fraction as determined by the Bohr equation. Addition of PEEP in prone position may lead to deterioration in V/Q mismatch although prone position itself can reduce the mismatch. Proper airway assessment and adequate planning, including preparation for awake fibreoptic intubation will be helpful if a difficult airway is anticipated as in our case.

To summarise, prone surgery in a patient with anteriorly protruding chest is a challenge to the anaesthesiologist. Performing the surgery in two stages and adopting measures to improve chest wall elasticity can help in reducing V/Q mismatch. Avoiding tranexamic acid and using Mayfield pin fixator instead of headrest can prevent POVL in high-risk patients.

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**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.
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Tency Babu, Preetha Chandran
Department of Anaesthesiology, Aster MIMS, Kozhikode, Kerala, India

Address for correspondence:
Dr. Tency Babu,
Department of Anaesthesiology, Aster MIMS, Govindapuram P.O., Kozhikode, 673 016, Kerala, India.
E-mail: tencybabu01@gmail.com

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