Anaesthesia Management for Edward's Syndrome (Trisomy 18)

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Dear Editor,

Edward's syndrome is the most common autosomal trisomy after Down syndrome (1). It is a syndrome in which multiple malformations such as structural heart defects, microcephaly, micrognathia, low-set ears, limbs, gastrointestinal and genitourinary system defects are observed (1). Death often occurs in the first year of life because of reasons such as severe heart defects, central apnoea, aspiration, respiratory failure and upper airway obstruction, found in more than 90% of patients (1, 2). Both serious heart problems and the possibility of a difficult airway due to micrognathia makes anaesthetic management special for patients to undergo surgery. There are few reports in the literature regarding the anaesthetic management of these patients.

A 2-month-old baby boy weighing 3030 g was evaluated for intervention because of a right inguinal hernia. It was found out that he had perimembraneous outlet ventricular septal defect (VSD), atrial septal defect (ASD), patent ductus arteriosus (PDA), tricuspid valve malformation, pulmonary hypertension, horseshoe kidney and, in the cranial system, ventriculomegaly and mega cisterna magna. Physical examination revealed that the baby with microphthalmia, micrognathia, short neck and ‘club foot’ deformity in lower extremities was low-weighted and apathetic (Figure 1). Findings of laboratory tests were normal. It was decided to place a laryngeal mask without using muscle relaxant in the patient in whom routine monitoring was conducted. Number 1 laryngeal mask was placed on the patient, who received 3 mg kg⁻¹ thiopental sodium and 2%–3% concentration of sevoflurane intravenously in the induction. In the anaesthesia maintenance, 50%/50% oxygen/nitrogen mixture was applied with 1%–2% concentration of sevoflurane. In the patient who remained haemodynamically stable, the intervention was completed smoothly, and no perioperative complications developed.

Edwards’ syndrome is a complex syndrome for anaesthesiologists, with many aspects. In these patients, in whom cardiac malformations are at the forefront, it is important to prepare an anaesthesia plan that will not affect the haemodynamics. Because Edwards’ syndrome may cause muscle rigidity, it is stated that succinylcholine should be avoided in cases where the use of muscle relaxants is required (3). Considering the possibility of difficult intubation that may occur because of micrognathia, we believe that it would be appropriate to use the supraglottic airway tools that require the use of lower doses of anaesthetic drugs in appropriate cases and cause less haemodynamic response.
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References

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