

Non-operating room anaesthesia for a child with Beckwith Wiedemann syndrome with a history of congenital laryngomalacia

Sir,

We report a 5-year-old boy with features of Beckwith Wiedemann syndrome (BWS) with a history of neonatal hypoglycaemia, stridor (congenital laryngomalacia), atrial septal defect, patent ductus arteriosus, undescended testis, disproportionate head growth, lethargy, delayed developmental milestones,

hydrocephalus with ventriculoperitoneal shunt surgery at 6 months of age, seizures, hypothyroidism, and left hemi-megalencephaly who presented for magnetic resonance imaging (MRI) under anaesthesia. He was on antiepileptics and thyroxine supplementation for the past 4 years, and his mother presently complained of noisy breathing and occasional stridor during sleep in the supine position which improved in the lateral position. The examination revealed asymmetric feet and hands with left-sided hemihypertrophy [Figure 1]. His thyroid profile and recent 2D echocardiography were normal. The growth hormone levels were elevated -3.19 ng/mL ($0.1-2.1$ ng/mL).

On the day of the MRI, the child fasted 6 h for solids and 2 h for clear fluids, had taken oral antiepileptics



Figure 1: Hemihypertrophy of the left side of the face (a) Anteroposterior view (b) Lateral view

and thyroxine. After attaching monitors such as electrocardiogram, pulse oximeter, and non-invasive blood pressure, anaesthesia was induced with a graded increase in sevoflurane concentration and oxygen while maintaining spontaneous ventilation with the child in the lateral position, and intravenous access was secured after induction. The anaesthesia was further deepened with 1 mg/kg propofol, and the airway was secured with i-gel supraglottic device (size 2). During the procedure, the anaesthesia was maintained with sevoflurane [0.8 minimum alveolar concentration (MAC)] in oxygen and air mixture with controlled ventilation. The intraoperative random blood sugar was normal. The procedure lasted for 45 min. After the procedure, i-gel was removed as the child became fully awake and was nursed in the lateral position. The child was discharged after 6 h of observation as per day-care protocol.

Anaesthetic challenges in BWS are abnormal airway anatomy, cardiac anomalies, and hypoglycaemia. Increasing the depth of anaesthesia may result in tongue fall, leading to upper airway obstruction, which is a challenging scenario during inhalational induction.^[1] Anticipation of difficulty, judicious use of airway aids, and graded inhalational induction result in successful airway management. These patients are prone to develop sleep-disordered breathing problems such as snoring/abnormal noises during sleep, postnatal respiratory distress, and obstructive sleep apnoea due to macroglossia, laryngomalacia, or tracheomalacia which can persist up to 15 years of age.^[2]

The cause for neonatal stridor is laryngomalacia from the collapse of supralaryngeal structures during inspiration which subsides by 1 to 2 years of age.^[3] However, the unmasking of laryngomalacia in a 6-year-old child with a history of neonatal

laryngomalacia (which had completely resolved after 4.5 years of age), has been previously reported during induction of anaesthesia wherein the child developed inspiratory stridor and desaturated to 92% and had bradycardia immediately after spontaneous induction of anaesthesia with 8% sevoflurane and oxygen.^[4]

Various complications such as intraprocedural unplanned intubation, pulmonary aspiration, procedure cancellation due to difficulty in maintaining airway, cough, and secretions complicating airway management have been reported during sedation for MRI procedures with pentobarbital and propofol.^[5,6] Hence, we decided to avoid the procedure under sedation and preferred general anaesthesia using i-gel after a graded increase in sevoflurane induction for the same. In addition, i-gel is superior to other supraglottic airway devices as it does not produce artifacts with MRI.^[7]

Anaesthetic challenges included managing a syndromic child having congenital laryngomalacia with persistent stridor for day-care anaesthesia in a remote location. We suggest that general anaesthesia with i-gel after slow-graded inhalational induction is the key for facilitating safe MRI in such a group of patients.

Although there are case reports regarding anaesthetic management of patients with BWS for surgery, ours is probably the first describing the safe management of such patients for procedures in remote locations.

Declaration of patient consent

Written informed consent was obtained from the mother of the patient whose details are described here.

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Conflicts of interest

There are no conflicts of interest.

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