Bifid epiglottis, high-arched palate, and mental disorder in a patient with Pallister-Hall syndrome

Sir,

Pallister–Hall syndrome is a rare autosomal recessive disease first described in 1980 by Philip Pallister and Judith Hall.[1] Craniofacial anomalies include bifid epiglottis, high-arched palate, and external ear anomalies.[1,2] In addition, polydactyly and hypothalamic hamartoma causing hypopituitarism and neurological disorders are also commonly seen.[3] Imperforate anus and renal anomalies occur frequently.

We report the anaesthetic management of a girl with Pallister–Hall syndrome under general anaesthesia. A 4-year-old female, 98.4 cm in height and 21.1 kg in weight, body mass index 21.8, with Pallister–Hall syndrome, was scheduled for dental treatment under general anaesthesia. She was diagnosed to have localized epilepsy due to hypothalamic hamartoma for which she was receiving sodium valproate 200 mg. With this medication, her epilepsy was partially under control and she continued to have minor seizure activity, a few times a day. Her neurological status was poor with severe mental retardation. In addition, we could not communicate with her.

Her clinical features included a high-arched palate and preauricular tags. The clinical assessment of her airway was difficult due to uncooperative behavior [Figure 1]. In addition, she had renal abnormalities and hand polydactyly. No abnormal findings were observed on chest X-ray and laboratory data.

She was brought to the operating room after peripheral intravenous access was obtained. Anaesthesia was induced with fentanyl 50 μg, atropine 0.2 mg, and propofol 40 mg after preoxygenation and with monitoring of SpO_2, electrocardiography, blood pressure, and heart rate. After the loss of consciousness, mask ventilation without an oral airway was easy. Then, rocuronium 12 mg was administered. During intubation using a 5.0-mm uncuffed nasotracheal tube by direct laryngoscopy, a bifid epiglottis was

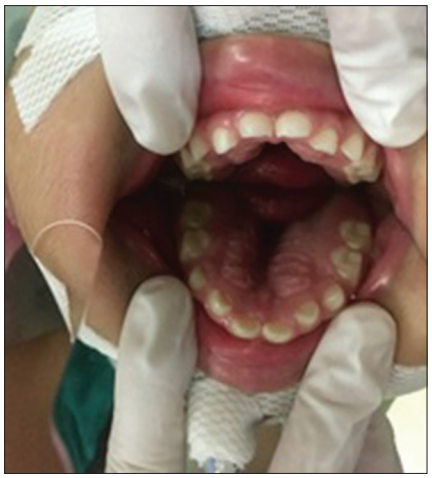

Figure 1: High-arched palate

noted [Figure 2]. Anaesthesia was maintained with end-tidal isoflurane 0.9–1.8 in air and oxygen. Haemodynamic parameters were stable, bispectral index value was 54–68, spectral edge frequency was 16–23 Hz, signal quality index was 95%, and $EtCO_2$ was 35–45 mmHg. Dental treatment was completed in 109 min without any surgical problems. There was minimal blood loss during operation, and she received a total of 303 mL lactated Ringer's solution with 1% glucose. Urine volume was 80 mL. After extubation, her postoperative stay was uneventful, and she left the hospital 1 day later.

Pallister–Hall syndrome is an autosomal dominant disorder characterized by varying combinations of a spectrum of abnormalities. However, there was rare case report about anaesthetic management in literature.[2] The importance of anaesthetic management was safe airway management and to avoid epilepsy.

In dental treatment, nasal intubation is usually performed, as it provides unrestricted access to the mouth and facilitates insertion of instrumentation. Airway management for patient with craniofacial disorders poses many challenges.[4] In this case, difficult intubation was especially anticipated because of the presence of high-arched palate and the preoperative airway assessment. During the induction of general anaesthesia, difficult airway cart and expertise should be made available and checked. Intubating stylet, video laryngoscope, fiberscope, or tube exchanger should be available. Airway management depends on evaluation of the abnormalities in each case, and modifications of technique might be needed to decrease the risk of airway complications.

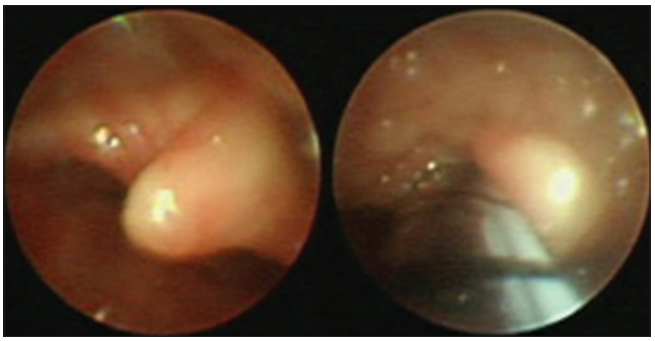

Figure 2: Bifid epiglottis before intubation (left side) and during intubation (right side)

In this case, we found a bifid epiglottis, which is a congenital malformation defined as a midline cleft of the epiglottis. It presents as a component of multiple anomalies as a syndromic constituent of malformation syndromes. It was reported in 40% of patients with Pallister–Hall syndrome.[2] It is typically asymptomatic but may be associated with stridor chronic, aspiration, and rarely airway obstruction.

Epilepsy is a serious neurological disorder during general anaesthesia. It can be seen as a result of imbalance between excitatory and inhibitory neuronal activity.[5] Appropriate perioperative management of antiepileptic drugs therapy is important to maintain epilepsy control. Anaesthesiologists need to be aware of the pharmacological properties of commonly used drugs. In patients with a history of status epilepticus, regular medications should be taken on the morning of general anaesthesia.

Isoflurane has well-characterized anticonvulsant properties of refractory status epilepticus.[5] In addition, propofol was administered to avoid epileptic activity during the induction of anaesthesia. As hyperventilation might decrease cerebral blood flow leading to epilepsy, we had maintained $EtCO_2$ within normal level.

We concluded that preparation for a difficult intubation was essential, although in previous reports and in our case, a difficult airway was not encountered. In addition, good control of epilepsy would contribute in decreasing perioperative morbidity.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be

reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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