Unique airway finding in a case of Pfeiffer syndrome and its management

Sir,

Pfeiffer syndrome is a rare craniofacial anomaly with an incidence of 1 in 100,000 births worldwide. It is rare in the Asian population, with only a few cases reported. The diagnosis of Pfeiffer syndrome is based on the presence of craniosynostosis and abnormal thumbs and/or first toes. It occurs due to mutations of fibroblast growth factor receptor (FGFR) genes 1 and 2.\(^1,2\)

A 42 kg, 22-year-old woman, a diagnosed case of Pfeiffer syndrome, was posted for septoplasty for correction of Deviated Nasal Septum (DNS). She gave a history of difficulty in breathing and had exophthalmos, midfacial hypoplasia, protruding mandible, small nose, broad thumbs, and big toe with deviation away from other digits. Her brother too had similar features.

Difficult Bag Mask Ventilation and intubation was anticipated and a difficult airway cart was prepared. Inhalation induction without muscle relaxation was planned. In the operation theatre, a 20 G IV line was secured, standard monitors were attached and 1 mg midazolam and 40 µg fentanyl were given intravenously. After preoxygenation for 3 min, anesthesia was induced with incremental doses of sevoflurane in 100% oxygen. Under deep anesthesia, direct laryngoscopy revealed a Cormack and Lehane grade IV view, which improved to grade III after optimum external laryngeal manipulation. A membranous laryngeal web was also seen posteriorly. Tracheal intubation with a 6.5 mm ID cuffed endotracheal tube (ETT) mounted on a stylet was not successful. A bougie was inserted (although with some difficulty) and a 6.0 mm ID ETT was successfully rail-roaded over it.

 Syndromes with craniofacial anomalies, such as Pfeiffer’s, are very rare and can be a challenge to anesthesiologists. These can be managed successfully with meticulous planning and execution.

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Figure 1: A patient with Pfeiffer syndrome
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Leaks in the vaporizer unit: Still a possibility

Sir,
Modern day vaporizers are reliable and come with many safety features. We report a case of an unusual leak from the vaporizer unit. The incident occurred while conducting anesthesia in a patient with mandibular fracture, posted for open reduction and internal fixation, on replacement of the halothane vaporizer with isoflurane vaporizer. After replacement, there was sudden under-filling of the reservoir bag after turning the control dial on. The flow meters showed no change in the flow, which meant there was a leak. The connections within the circuit, between the circuit, and the common gas outlet and filler cap of the vaporizer were checked out for leaks, but no leaks were detected. On checking the vaporizer mount, a needle was found between mounting system and the vaporizer, which was causing the leak. The leak was then rectified.

Detachable mounting systems, like Selectatec, permit movement and change of the vaporizer unit. These consist of a pair of port valves for the vaporizer. Each vaporizer has a special mounting bracket that fits over the nipples. The weight of the vaporizer and an O-ring around each port valve create a seal between the mounting system and the vaporizer. [1]

In case of a leak in the vaporizer, the anesthesia delivery machine can function normally if the vaporizer remains switched off. On switching on the vaporizer, the fresh gas flow is reduced with little or no vapor. A common cause of leak is failure to replace the filler cap or tighten it. [2] If an incorrect cap is used on a filler device, leak may occur. [3] Another common cause of the leak is the absent or damaged O-ring. [1,4] Other locations for leaks include the selector valve, mounting mechanism, interlock device, vaporizer inlet, or outlet connections. [1,4]

In our case, though a needle was present between the mounting system and vaporizer [Figure 1], it was possible to lock the locking lever and the control dial could be turned on. This led to leakage of fresh gas flow and under-filling of the reservoir bag. Consequences of leaks include alteration in the flow and composition of inspiratory gas and pollution of the operating room air. The former can cause hypoxia, re-breathing or awareness in a patient if not identified and rectified.

In conclusion, detachable mounts are convenient but are a potential source of leak. We recommend that utmost care be taken during dismounting and remounting of vaporizers to avoid damage to them and prevent leaks.

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Figure 1: Needle causing the leak

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