Anaesthesia for ventriculoperitoneal shunt surgery in a craniovertebral junction anomaly patient with arrested hydrocephalus: Stability is safety

Sir,

We report the anaesthetic management of a 23-year-old woman for ventriculoperitoneal (VP) shunt surgery who presented with history of increasing head size from 2 months of age, weakness of right upper and lower limbs from her fifth birthday. She also suffered from on and off headache since 10 years which had worsened since 2 months. Her Glasgow Coma Score was 15, with right upper and lower limb power of grade 3 and 4 respectively, without any sensory, cerebellar or cranial nerve involvement. She had bilateral genu valgum deformity and a restrictive pattern in pulmonary function tests. Magnetic resonance imaging showed fracture of odontoid tip (Osodontoideum), C1 anterior arch with C2 retropulsion impinging the cord, causing severe myelomalacia and syrinx from C2 to C6 level with fourth ventricular outflow tract obstruction [Figure 1].

She was adequately fasted on the day of surgery and received nebulisation with 4% lignocaine in the preoperative room. In the operation theatre after connecting standard monitors, intravenous (iv) glycopyrrolate 200 µg, fentanyl 60 µg and a loading dose of dexmedetomidine at 0.5 µg/kg over 10 min (continued with an infusion of 0.3 µg/kg/h, until intubation) followed by a transtracheal block with 2 ml of 4% lignocaine, were given. Airway was secured with awake fibreoptic bronchoscopic intubation using a 7 mm internal diameter endotracheal tube. Anaesthesia was induced with iv propofol 80 mg and atracurium 25 mg and maintained with sevoflurane (0.8 minimum alveolar concentration) in oxygen-air mixture and atracurium boluses under controlled ventilation. Patient was positioned supine with head turned to the left, which was stabilised with Mayfield 3 pin head clamp (after an infiltration of 1.5 ml of 2% lignocaine at each pin site), which remained throughout the procedure [Figure 2]. Intravenous boluses of fentanyl 50 µg and propofol 30 mg were given before tunnelling. The procedure was uneventful, head clamp was removed, neck collar placed and the patient was reversed and extubated once fully awake. Postoperative neurological status remained the same as preoperative.

Osodontoideum is a rare type of craniovertebral junction (CVJ) anomaly in which upward migration of a separated ossicle of hypoplastic odontoid process results in atlantoaxial insufficiency (AAI).[1] This AAI can further cause spinal canal compression, restricting cerebrospinal fluid circulation leading to hydrocephalus[1,2] which sometimes gets arrested. The overall process leads to restrictive lung disease. Arrested hydrocephalus often has a stable period with subacute neurologic decline, rarely progressing to further ventricular enlargement. Occasionally sudden and fatal decompensation of arrested hydrocephalus occurs due to stretching of the axons which control the cardiorespiratory centres.[3] Odontoid process

Figure 1: MRI image showing fractured odontoid tip, C1 anterior arch with C2 retropulsion, impinging the cord, causing severe myelomalacia and syrinx at C2-C6 level with 4th ventricular outflow tract obstruction

Figure 2: Patient positioned for VP shunt surgery with head fixed using Mayfield head clamp
anomalies can be associated with trisomy 21 and Klippel-Feil malformations\(^1\) presenting with difficult airway, making anaesthetic management more challenging.

Awake fibreoptic bronchoscopy (FOB) minimises neck movements during intubation.\(^2\) Anxiety, hypercarbia and haemodynamic fluctuations can raise the intracranial pressure further.\(^3\) Dexmedetomidine sedation during FOB has a favourable profile in this scenario, by reducing anxiety without significantly affecting respiration or haemodynamics\(^(b)\) especially when preoperative use of anxiolytics is detrimental; nevertheless, dexmedetomidine is often used as a sedative and analgesic adjunct intraoperatively for airway procedures.\(^1\) Most commonly used head ring for head and neck positioning during VP shunt procedure may not provide a stable neck position especially during tunnelling and can worsen the CVJ instability and neurologic deterioration postoperatively. Mayfield head clamp is rarely used in VP shunt surgery, but its use here will solve this issue. Haemodynamic fluctuations during head clamping and tunnelling can be suppressed with local lignocaine infiltration and boluses of opioid or propofol, respectively.\(^2\) Since these patients have restrictive lung disease, extubation should be performed carefully when fully awake after complete reversal from muscle relaxant. Placing a collar prior to extubation ensures neck stability further.

Meticulous care during airway manipulation and use of a Mayfield clamp for head fixation as in our case would prevent postoperative neurological deficits and facilitate safe anaesthesia. To the best of our knowledge, only one case report on anaesthetic management in such patients (for occipito-cervicalfusion surgery) exists; however, ours is probably the first, describing anaesthesia for such patients undergoing VP shunt surgery using Mayfield clamp which is unique.

**Declaration of patient consent**
Written informed consent was obtained from the mother of the patient who’s details are described here.

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**Conflicts of interest**
There are no conflicts of interest.