Challenges of repetitive sedation in a 16-month old child undergoing proton beam therapy

Sir,

Anaesthesia for proton beam therapy (PBT) demands excellence in paediatric anaesthesia[1] and an understanding of functioning of PBT and the limitations of working in a shared workspace.

A 16-month-old child with grade-3 anaplastic ependymoma weighing 8.5 kg with delayed motor milestones and a large head due to hydrocephalus was referred to us. He had undergone two craniotomies, a shunt revision and had completed five cycles of chemotherapy. He was on levetiracetam and had moderate iron deficiency anaemia with a haemoglobin of 9.2 g/dL.

Planning phase involved magnetic resonance imaging (MRI), immobilisation device making and computed tomography (CT) and lasted 180 min. The amount
of mouth opening and head extension to maintain spontaneous respiration without use of artificial airway, the position of spine and limbs were confirmed during planning phase as this needed to be reproduced during therapy [Figures 1 and 2]. Therapy phase involved 33 sessions and 2 quality assurance CT and was done 5 days a week. Each session lasted about 35 min (range; 20–80). 1–2 mg/kg of propofol administered in 1 mL incremental boluses to achieve immobilisation followed by a maintenance dose of 10 mg/kg/h was used. No repeat bolus was required during therapy. No adverse anaesthesia events were observed.

PBT sedations are associated with challenges of non-operating room anaesthesia (NORA) but are different from conventional radiotherapy with regard to the younger age, longer in room time and stringent immobilisation techniques necessitating deep sedation. A lighter plane may result in incomplete mouth opening and airway obstruction when the mask is applied. Incompletely relaxed limbs and spine will need repositioning. Irradiation to head and neck can cause airway and oesophageal oedema, increased secretions and hyper-reactive airway, coughing and laryngospasm.

Hypothermia from exposure (cling wraps were used to cover the child), hypoglycaemia due to unplanned delay arising from technical issues (dextrose containing fluids were used), the physiological effects of radiation like anaemia and weight loss (haemoglobin was 7.2 g/dl and weight was 7.6 kg towards the end of therapy) that affect anaesthesia outcomes and ensuring feeding and discharge within an hour daily were other challenges.

Though multicentric trials suggest safety of short exposure to general anaesthesia, there is no literature on the effects of successive sedation on neural development. Neurosurgery, chemotherapy, radiation and anticonvulsant medications also affect cognition. Guidelines on the age for the first anaesthetic application, importance of duration, number or interval between two consecutive anaesthesia applications are ill defined. Choosing the least harmful anaesthesia technique for safe and effective sedation daily in a NORA setup for a very small child with cancer is the real challenge in PBT.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the legal guardian has given his consent for images and other clinical information to be reported in the journal. The guardian understands that names and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

**Financial support and sponsorship**

Nil.

**Conflicts of interest**

There are no conflicts of interest.

---

*Indumathi Dalvam, Anand Murugesan, Pankaj K Pande, Rakesh Jalali*

Department of Anaesthesia and Pain Management, *Clinical Research and Radiation Oncology, Neuro-Oncology Cancer Management Team, Apollo Proton Cancer Centre, Chennai, Tamil Nadu, India*
Sir,

Pompe disease is an autosomal recessive type II glycogen storage disorder affecting 1 in 40,000 live births. It is caused by the deficiency of lysosomal α-glucosidase (GAA) enzyme that degrades lysosomal bound glycogen to glucose. [1]

We present the anaesthetic management of a 21-year-old primigravida at 37 weeks of gestation, a known case of juvenile Pompe’s disease, posted for elective caesarean section. Her symptoms started at 10 years when she noticed difficulty in getting up from squatting position. The diagnosis was confirmed by leukocyte enzyme analysis and muscle biopsy. She was started on enzyme replacement therapy (ERT) alglucosidase alfa 20mg/kg intravenous infusion over 5h every 2 weeks. She conceived spontaneously and continued to get ERT throughout her pregnancy. At 34 weeks gestation, she was admitted for safe confinement and was started on oral ubiquinone 90mg once daily and injection enoxaparin 0.4 mg subcutaneously once daily. Blood investigations and echocardiogram were normal. Pulmonary function tests in standing and sitting position showed severe restriction [Table 1 and Figure 1].

For caesarean section, subarachnoid block was given. She delivered a live healthy female baby of 2.8 kg. After delivery, oxytocin infusion was started. The procedure was uneventful. Postoperative analgesia was provided with ultrasound-guided bilateral transversus abdominis plane block with 30 ml of 0.1% ropivacaine and intravenous 1g paracetamol 8th hourly. She had an uneventful postoperative period and received one more dose of ERT infusion. She was discharged on the 5th postoperative day.

Based on the age of onset, Pompe disease is classified into infantile and late-onset. Infantile form is severe and presents with failure to thrive and feeding difficulties. Late-onset Pompe disease can present at any age. They can also have kyphoscoliosis. [2]

Proximal muscles of the lower limb and paraspinal trunk muscles are affected initially followed by diaphragm. Pregnancy is risky in these patients as the mass effect of the uterus could further compromise the respiratory function. Elective caesarean section is usually preferred in these patients. Enzyme replacement therapy (ERT) with alglucosidase alfa, though expensive is found to be effective and safe.

---

1. Sen I, Dave N, Bhardwaj N, Juwarkar C, Beegum S. Specialised training in paediatric anaesthesia: Need of the hour. Indian J Anaesth 2021;65:17-22.
2. Maddirala S, Theagrajan A. Non-operating room anaesthesia in children. Indian J Anaesth 2019;63:754-62.
3. Davidson AJ, Disma N, de Graaff JC, Withington DE, Dorris L, Bell G, et al. GAS consortium. Neurodevelopmental outcome at 2 years of age after general anaesthesia and awake-regional anaesthesia in infancy (GAS): An international multicentre, randomised controlled trial. Lancet 2016;387:239-50.
4. Warner DO, Zaccariello MJ, Katusic SK, Schroeder DR, Hanson AC, Schulte PJ, et al. Neuropsychological and behavioral outcomes after exposure of young children to procedures requiring general anaesthesia: The Mayo Anaesthesia Safety in Kids (MASK) Study. Anaesthesiology 2018;129:89-105.
5. Robinson KE, Kuttesch JF, Champion JE, Andreotti CF, Hipp DW, Bettis A, et al. A quantitative meta-analysis of neurocognitive sequelae in survivors of pediatric brain tumors. Pediatr Blood Cancer 2010;55:525-31.