Central core disease with scoliosis for congenital hip dislocation surgery: An anaesthetic demur

Sir,

Central core disease (CCD), an inherited disorder, presents with hypotonia, proximal weakness, scoliosis, hip dislocation and susceptibility to malignant hyperthermia (MH).[1-3]

Patients with CCD have been reported to experience a variety of life-threatening complications during and after general anaesthesia.

We report a case of CCD (proven by muscle biopsy) and very severe kyphoscoliosis with a thoracic and thoracolumbar component and Cobb's angle of 90 each [Figure 1], scheduled for right hip surgery. The patient was a 3-year-old female and weighed 11 kg. There was no history of previous anaesthetic exposure or anaesthetic problems in the family. Informed consent was obtained from the parents.

On the day of surgery, the patient was premedicated with oral midazolam 0.75 mg/kg, 30 min before the procedure. Intravenous access was secured. Glycopyrrolate 4 mcg/kg and ketamine 1 mg/kg were given intravenously. ECG, NIBP, SpO2, $EtCO_2$, temperature and urine output monitoring was used. Maintenance of anaesthesia was by intravenous infusion of propofol 6–8 mg/kg/hour with fentanyl 1 mcg/kg/hour. Continuous sidestream capnography was performed while the face mask was in place. The patient received spontaneous ventilation with Jackson–Rees (JR) circuit.[4] The JR circuit was connected directly to an oxygen humidifier through extension tubes. This bypassed the anaesthesia machine and its elaborate cleanup of traces of volatile agents from all components.

The $EtCO_2$, SpO_2 and the movement of the JR circuit bag indicated adequacy of ventilation. With the patient in lateral position, under all aseptic precautions, the L2–L3 epidural space was accessed with an 18 G Tuohy needle using loss of resistance to saline at a depth of 2.5 cm. An epidural catheter was inserted and secured at 8 cm at the skin. Epidural catheter tip was situated at L1 and L2. The exact placement of the epidural catheter was confirmed fluoroscopically, in anteroposterior view using [Figure 1], 1 ml epidural

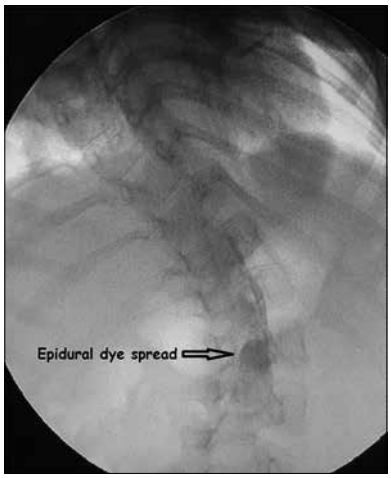

Figure 1: Carm (anteroposteriorview) of spine showing proper placement of the epidural catheter using contrast dye

non-ionic contrast dye (Omnipaque™ GE Healthcare, Cork, Ireland).

Initial epidural bolus test dose of 2 ml of 2% lignocaine with 1: 200,000 adrenaline was given after negative aspiration for blood and cerebrospinal fluid, followed by 5 ml (0.5 ml/kg) of 0.2% ropivacaine. This was repeated every 2 h. There was no change in the baseline heart rate and blood pressure to surgical stimuli which further confirmed the adequacy of the epidural blockade. Stable haemodynamics were maintained throughout the procedure and the intraoperative course was uneventful. Around 20 min before the end of procedure, propofol and fentanyl infusions were stopped, resulting in smooth emergence. The patient was awake, conscious, responding to verbal commands and had sufficient pain relief. An epidural infusion of 0.2% ropivacaine at 0.4 mg/kg/hour was started post-operatively. This provided post-operative analgesia for 72 h.

MH susceptible patients do pose a serious challenge. In our patient, it was further compounded by the presence of severe kyphoscoliosis. Despite the presence of a difficult spine, the technique of combined epidural and total intravenous anaesthesia avoided the administration of medications known to be triggers for MH. In addition, epidural analgesia is an excellent method for perioperative pain management in a child with severe restrictive lung disease.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

Vrushali C Ponde, Vinit Vinod Bedekar, Ashok N Johari[1], Shalin K Maheshwari[1]

Department Of Anaesthesiology, Children's Anaesthesia Services and Surya Children Hospital, [1]Department of Pediatric Orthopedic Surgery, Enable - International Centre for Pediatric Musculoskeletal Care, Mumbai, Maharashtra, India

Address for correspondence:
Dr. Vinit Vinod Bedekar,
B-41/201, Yogi Nagar, Yogi Saraswati CHS., Eksar Road,
Borivali (West), Mumbai - 400 091, Maharashtra, India.
E-mail: dr.vinitbedekar@gmail.com

REFERENCES

1. Munster T. Anaesthesia recommendations for patients suffering from Central Core Disease. Available from: https://www.orpha.net/data/patho/Pro/en/Central_Core_EN.pdf. [Last accessed on 2017 Dec 17].
2. Jimi N, Izumi K, Sumiyoshi R, Mizuno K. Anesthetic management of a patient with central core disease for scoliosis surgery. Masui 2016; 65:649-51.
3. Eng GD, Epstein BS, Engel WK, McKay DW, McKay R. Malignant hyperthermia and central core disease in a child with congenital dislocating hips. Arch Neurol 1978; 35:189-97.
4. Ponde V. Surgical mask used as a harness. PaediatrAnaesth 2006; 16:601.

Access this article online	
Quick response code	Website: www.ijaweb.org
	DOI: 10.4103/ija.IJA_570_17

How to cite this article: Ponde VC, Bedekar VV, Johari AN, Maheshwari SK. Central core disease with scoliosis for congenital hip dislocation surgery: An anaesthetic demur. Indian J Anaesth 2018;62:84-5.