Anaesthetic challenges in a child with sickle-cell disease and congenital heart block

Sir,

We present a case report of anaesthetic management of a 12-year-old boy with sickle-cell disease (SCD) and congenital heart block (CHB) with pacemaker [Figure 1], undergoing laparoscopic splenectomy.

The child had suffered sickle-cell crisis and pacemaker malfunction a month back. His preoperative investigations were normal and weight was 30 kg. He received intravenous (IV) fluids during fasting period, pneumococcal vaccine, antibiotic prophylaxis and cefotaxime 500 g IV. The cardiologist changed the mode of pacemaker to asynchronous (fixed rate). Standard

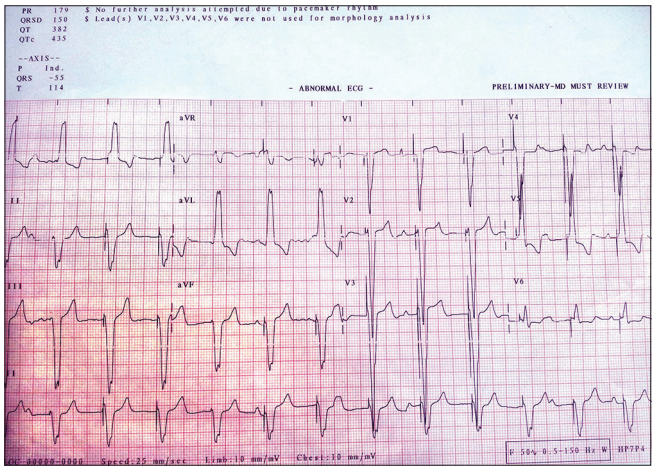

Figure 1: 12-lead electrocardiography of the patient

American Society of Anesthesiologists monitoring, urine output and temperature monitoring were used.

Morphine 4.5 mg IV, thiopentone 150 mg IV and atracurium 0.5 mg/kg IV were used for induction and intubation. Anaesthesia was maintained with isoflurane (MAC: 1.0), air and oxygen (FiO_2: 0.5). Pneumoperitoneum was created with CO_2 and intra-abdominal pressure was kept around 8–10 mmHg. The surgeon used bipolar cautery, blood loss was approximately 800 ml and 1 unit packed red cell was transfused. The patient was given fluid at 60 ml/h intra-operatively as he had been kept hydrated preoperatively. For post-operative analgesia, paracetamol 500 mg IV was given and skin incision was infiltrated with 12 ml 0.2% ropivacaine. Neuromuscular blockade was reversed and trachea was extubated. The patient remained haemodynamically stable, normothermic and had adequate urine output. Post-operative analgesia was maintained with paracetamol 500 mg IV 8th hourly and fentanyl boluses of 30 μg as rescue analgesic whenever visual analogue scale exceeded 3. The child was discharged on the 7th post-operative day after uneventful course.

CHB (incidence 1 in 22,000 live births) with no structural abnormality and when diagnosis is established beyond neonatal period has better survival.[1] Intraoperatively, pacemaker mode is advised to be changed to asynchronous, and bipolar electrocautery is to be used. The availability of manual pacing should be ensured to manage pacemaker malfunction.[2]

SCD is the disorder of beta-globin chain characterised by haemolytic anaemia, intermittent vascular occlusion, pulmonary compromise and multi-organ damage. Electrocardiogram changes are non-specific

and first-degree heart block may be found.[3,4] Not usually associated with SCD, CHB in our patient is perhaps an additional finding.

Splenectomy is recommended in children older than 2 years or after one major or two minor episodes of splenic sequestration crises.[3,4] When done laparoscopically, the duration of surgery increases, but hospital stay is decreased.[3] Any major surgery is associated with 7% mortality. Preoperatively, the history of episodes of painful crises must be sought.[4] Pre-operative examination should be thorough as SCD involves multiple organ systems. Neurological evaluation rules out the previous cerebrovascular accident as ischaemic infarcts are common in anaemic children.[3] Kidneys may get involved during advanced disease. Presence of rib infarcts can lead to hypoventilation. Perioperative cardiopulmonary manifestations such as acute chest syndrome usually remain undetected in young children.[3,4]

As dehydration precipitates RBC sickling and occlusion of microvasculature at a level of precapillary sphincters, perioperative hydration must be ensured.[3,4] There is a controversy regarding blood transfusion, benefit being dilution of haemoglobin S and disadvantage being triggering of sickling.[3,4] Other factors leading to sickling crises are vascular stasis, hypoxia, metabolic acidosis, hypothermia and presence of infection.[3] Previous case reports have emphasised the importance of preventing sickling crisis perioperatively.[5]

To conclude, perioperative anaesthetic management of children suffering from SCD and CHB needs meticulous pre-operative and intra-operative management and post-operative vigilance for preventing sickle-cell crisis and associated complications.

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Conflicts of interest
There are no conflicts of interest.

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