Sustained intraoperative bradycardia revealing Sengers syndrome

Sir,

Hypertrophic cardiomyopathies (HCM) are uncommon disorders in childhood.[1] They are often asymptomatic and can present as cardiac arrest. During preanaesthetic assessment, if bilateral congenital cataracts are present, the clinician should suspect Sengers Syndrome and search for an associated cardiomyopathy.

We report the case of a 6-month-old infant, born to a first-degree consanguineous marriage, weighing 6 kg and scheduled for congenital cataract surgery. Due to the delay in the management, surgery became urgent, and preanaesthetic consultation was carried out on the ophthalmology ward, 1 day before surgery. There were no particular functional signs, and clinical examination was essentially normal; in particular, there was no heart murmur or signs of heart failure. The child had a mild rhinopharyngitis without fever or rales. The anaesthetist judged the patient fit for anaesthesia without further investigations.

In the operating theatre, after induction of anaesthesia with sevoflurane, the infant received intravenous 2 μg/kg of fentanyl and 3 mg/kg of propofol. At the moment of intubation, incomplete laryngospasm occurred, so anaesthesia was deepened by administrating 30 mg of Propofol, which permitted an easy intubation. However, bradycardia (heart rate 55 bpm) occurred and persisted despite administration of atropine. This bradycardia was not preceded by significant desaturation, and $EtCO_2$ was at 8 mmHg. External cardiac massage was initiated and 100 μg of epinephrine IV administered, which resulted in restoration of an efficient circulation.

Surgical procedure was completed on the right eye within 15 min, and the patient was transferred

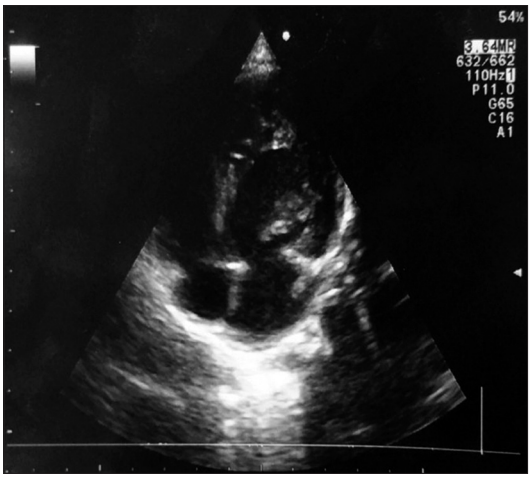

Figure 1: Apical four chamber ultrasound view showing an hypertrophied left ventricle for the age, with abnormal trabeculations

to the Intensive Care Unit. Transthoracic echocardiogram (TTE) showed a hypertrophied left ventricle (LV) for the age, with abnormal trabeculations [Figure 1], and apical hypokinesis of the LV. There was no obstruction to systolic ejection or associated congenital heart disease. The chest X-ray showed cardiomegaly [Figure 2].

The patient was administered low dose dobutamine and norepinephrine, which improved contractility without causing dynamic LV outflow tract obstruction. Extubation was carried out 2 h postoperatively, and noninvasive ventilation was applied. The patient improved, but a moderate lactic acidosis persisted. The association of bilateral congenital cataracts, hypertrophic cardiomyopathy and lactic acidosis characterises Sengers syndrome.

Pre-anaesthetic assessment of children is based generally on anamnestic data and clinical examination, usually without the need for further investigations.[2] During the neonatal period, even careful clinical examination detects only 45% of HCM.[3] It may remain silent and cause cardiac arrest in the operating room. The existence of a bilateral congenital cataracts, even in asymptomatic infants, should trigger the anaesthetist to order a TTE before anaesthesia.

Haemodynamic effects of propofol are exacerbated in the case of HCM. In fact, an impaired LV relaxation and compliance worsens the decrease of the stroke volume due to the drop of venous return. Myocardial hypertrophy increases the oxygen demand leading to myocardial ischaemia, in this context of decreased

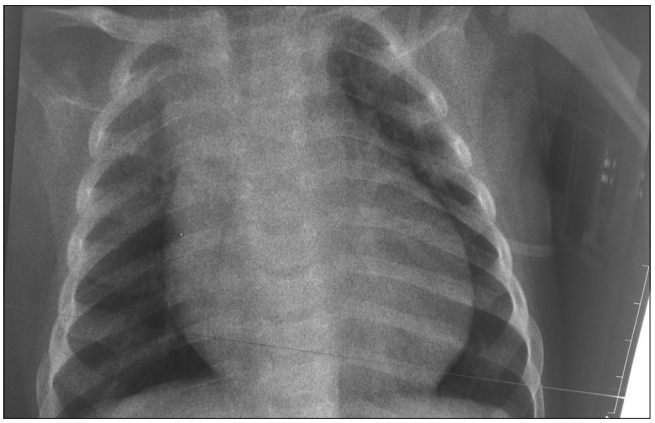

Figure 2: Chest X-ray showing cardiomegaly

coronary perfusion. However, rather than the choice of the hypnotic agent, it is how to use it which is decisive for haemodynamic stability.[4] Balanced anaesthesia is the reference in the management of patients with cardiomyopathy. In the case of a situation which requires deepening of anaesthesia such as laryngospasm, it makes more sense to use succinylcholine to allow intubation, instead of using propofol. Sevoflurane can be used cautiously and seems to be fairly well tolerated.[5] The use of sympathomimetic agents may intensify relaxation disorders and promote myocardial ischemia and dynamic obstruction.[3] In our patient, the use of low doses of dobutamine was supported by the presence of LV hypokinesis probably secondary to a myocardial stunning following the circulatory arrest.

Declaration of patient consent

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

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Conflicts of interest

There are no conflicts of interest.

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REFERENCES

1. Nugent AW, Daubeney PE, Chondros P, Carlin JB, Cheung M, Wilkinson LC, *et al.* The epidemiology of childhood cardiomyopathy in Australia. N Engl J Med 2003;348:1639-46.
2. Tay CL, Tan GM, Ng SB. Critical incidents in paediatric anaesthesia: An audit of 10000 anaesthetics in Singapore. Paediatr Anaesth 2001;11:711-8.
3. Maron MS, Olivotto I, Betocchi S, Casey SA, Lesser JR, Losi MA, *et al.* Effect of left ventricular outflow tract obstruction on clinical outcome in hypertrophic cardiomyopathy. N Engl J Med 2003;348:295-303.
4. Kaplanian S, Chambers NA. Anesthesia in a child with newly diagnosed hypertrophic cardiomyopathy. Paediatr Anaesth 2006;16:1080-3.
5. Ing RJ, Ames WA, Chambers NA. Paediatric cardiomyopathy and anaesthesia. Br J Anaesth 2012;108:4-12.

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