

Unfavourable outcome after uneventful anaesthesia and surgery in a child with Hurler syndrome

Sir,

Anaesthetic management of Hurler syndrome has focussed primarily on the airway and its associated complications. Associated cardiovascular lesions with perioperative evaluation and management have not been highlighted in detail. We report a sudden death on the 1st post-operative day in a child with Hurler syndrome who had undergone an uneventful surgery.

A 5-year-old, 13 kg female child diagnosed with Hurler syndrome was admitted for repair of recurrent umbilical hernia. She was receiving enzyme replacement therapy (ERT). She had anaesthesia exposure for umbilical hernia repair and ventriculoperitoneal shunt insertion at 2 and 4 years age, respectively.

There was history of snoring but no sleep apnoea. She had increased facial hair, hazy cornea, broad nose, large tongue and mental retardation [Figure 1]. Electrocardiogram and chest X-ray were normal. Echocardiography revealed normal chamber sizes, valves and biventricular function with asymmetrical interventricular septal hypertrophy.

After attaching monitors, anaesthesia was induced with 100% oxygen and gradual increase in sevoflurane concentration with facemask. Assisted ventilation was possible with the oral airway. Intravenous fentanyl 25 mcg was administered, and oral intubation was accomplished with stylet guided 5 mm ID uncuffed endotracheal tube. Anaesthesia was maintained with oxygen, air, sevoflurane and atracurium. Lumbar epidural catheter was inserted at L2/L3 intervertebral space, and 0.25% bupivacaine 5 ml bolus was administered. The total duration of surgery was 90 min, and trachea was extubated uneventfully.

Epidural analgesia with 0.1% bupivacaine at 3 ml/h was continued in the ward till next day morning. On the evening of 1st post-operative day, the nurse noticed



Figure 1: Child with Hurler syndrome before surgery

a sudden increase in perspiration followed by flat line on the monitor. Intubation and resuscitation were done according to standard protocol, but it was not possible to revive the child after prolonged cardiopulmonary resuscitation. The child's parent refused autopsy.

The most worrisome aspect of airway management has been solved with stem cell transplant, ERT and newer airway gadgets. Deposition of glycosaminoglycans on cardiovascular structures may manifest as valvular stenosis and regurgitation, cardiomyopathy, ventricular arrhythmias, heart block and coarctation of aorta. Narrowing of coronaries may result in myocardial infarction.^[1]

In our case, respiratory cause for sudden death was ruled out, as the child was conscious and responding few minutes back. Meanwhile, the child did not receive any sedatives or opioids. Local anaesthetic toxicity was excluded as the epidural was stopped in the morning. Hence, we suspected cardiovascular aetiology as a more likely cause for the sudden death.

Malignant ventricular arrhythmias, complete heart block and acute myocardial infarction, are the major causes for sudden cardiac death in Hurler syndrome. Most commonly, echocardiography is done to assess cardiac function in these children. However, it identifies valvular lesions, chamber size, cardiac contractility and regional wall motion abnormalities but is not an ideal method to assess coronary artery pathology.

Belani *et al.* have reported two intraoperative deaths in Hurler syndrome due to sudden ST-T changes and ventricular tachycardia progressing to cardiac arrest.^[2] The first child had a normal functional status with

worsening cardiac function in echocardiography. Autopsy showed occlusion of both coronaries. The second child had normal ECG, echocardiography and thallium scan. Later autopsy showed marked concentric fibrosis of both coronaries. Subsequently, coronary angiography and radio nucleotide scan were included in their pre-anaesthetic assessment of children with MPS scheduled for bone marrow transplant. In our case, child may have undiagnosed coronary pathology that lead to sudden death in the ward.

We suggest along with the detection of airway anomalies; cardiac assessment should also receive priority due to the high incidence of coronary artery involvement.^[3] However, the feasibility of coronary angiography and radio nucleotide scan in all patients with Hurler syndrome in our setting is debatable. Hence, perioperative care of these children should be done as if they are having coronary artery disease.

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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