Sir,

Hypertrophic cardiomyopathy (HCM) is not uncommon in pregnancy but apical hypertrophic cardiomyopathy (AHCM) is a rare variety.\textsuperscript{[1]} It is a benign, incidentally diagnosed condition with a 5% incidence.\textsuperscript{[2]} A 22-year-old G2P1L1 presented to the emergency department (ED) with palpitations for one month, angina, breathlessness on minimal exertion (New York Heart Association grade III), and progressive limb oedema for 2 weeks, oliguria for 2 days and occasional dry cough for a few months. She had a history of similar complaints in her previous pregnancy 2 years back at the 8th month for which caesarean section was done under GA and postoperatively ventilated. The patient was lost to follow up. Her records were lost. Her family history was notable for sudden cardiac death in her grandfather at a young age.

At admission, the patient weighed 45 kg with a height of 150 cm (body mass index-20kg/m\(^2\)). She had bilateral pedal edema, elevated jugular venous pressure, heart rate of 116 beats/minute, respiratory rate (RR) of 20/min, blood pressure (BP) of 116/80 mm Hg, peripheral oxygen saturation (SpO2) of 75% on room air and 95% on O2 by mask (6 litres), loud first heart sound with a grade 3/6 mid systolic murmur, soft second heart sound and fourth sound, Decreased breath sounds on the left side and bilateral basal crepitation. Electrocardiography (ECG) revealed T wave inversion in leads I, II, V2-V6 [Figure 1]. Echocardiography revealed Hypertrophic Obstructive Cardiomyopathy (HOCM) with severe biventricular hypertrophy. The patient was initially managed by the obstetrician as HOCM by fluids and intravenous morphine. However, her condition worsened and anaesthesiologist and cardiologist opinion was sought. Our bedside echocardiography revealed HCM with asymmetrical septal thickness, predominant apical thickening more than 4 cm, left ventricular outflow tract (LVOT) gradient less than 10 mm hg, Mild tricuspid regurgitation, right ventricular systolic pressure 45 mm Hg, and grade 3 diastolic dysfunction [Figure 2]. The patient worsened and she was taken for emergency lower segment caesarean section because of fetal distress.

Preoperatively, she was administered a stat dose of IV furosemide 20 mg, ranitidine, and metaclopramide. Pre-induction arterial line and ultrasound-guided central venous line was inserted in the right internal jugular vein. On table, BP-130/80 mm Hg, PR-130/min, RR- 24/min, end-tidal CO2 – 60 mmhg, central venous

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{figure1.png}
\caption{ECG: ECG shows deep T wave inversion in lead I, II and V2-V6 due to abnormal thickening of the cardiac apex and RVH and LVH strain pattern}
\end{figure}

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{figure2.png}
\caption{Top figure: Parasternal short axis view with LV hypertrophy with apical predominance. Bottom figure: Parasternal long axis view showing no signs of LVOT obstruction}
\end{figure}
Letters to Editor

Dear Editor,

Dr. Chitra Rajeswari Thangaswamy,
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was advised whereas, in HOCM, liberal fluid therapy with severe diastolic dysfunction, fluid restriction synergistic stimulation. Avoiding an increase in the contractility, and avoiding load, maintaining euvolemia, normal sinus rhythm, goals include avoiding tachycardia, reducing afterload, maintaining normal sinus rhythm, increasing contractility, and avoiding oedema, furosemide was given before delivery with fluid restriction. Treatment of refractory cases includes heart transplant and apical myomectomy.

AHCM (Yamaguchi Syndrome), a rare variety of HCM with autosomal dominant inheritance pattern has an E101K mutation in the alpha actin gene. Diagnostic parameters include asymmetrical left ventricular hypertrophy confined to apex, apical wall thickness ≥15 mm, ratio of maximal apical to posterior wall thickness ≥1.5, and ace of spade ventricular cavity. AHCM can manifest as myocardial infarction, embolic events, AF, ventricular fibrillation, and congestive heart failure. HCM presents due to diastolic dysfunction with well-preserved systolic function. The physiological changes during pregnancy worsen the diastolic dysfunction. Poor predictors for this patient included E/A >2 and CHF. ECG changes in Yamaguchi syndrome include deep symmetrical T wave inversion which contradicted her prior diagnosis of HOCM. Transthoracic echocardiography is a non-invasive modality for diagnosing AHCM, but is usually missed. Treatment goals include avoiding tachycardia, reducing afterload, maintaining euvoelma, normal sinus rhythm, avoiding an increase in the contractility, and avoiding sympathetic stimulation. Since she had presented with severe diastolic dysfunction, fluid restriction was advised whereas, in HOCM, liberal fluid therapy is required. Because of tachycardia, metoprolol was given decreasing diastolic dysfunction and improving ventricular filling in non-obstructive HCM. Induction was done in this manner to avoid haemodynamic fluctuations. To prevent the worsening of pulmonary oedema, furosemide was given before delivery with fluid restriction. Treatment of refractory cases includes heart transplant and apical myomectomy.

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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5. Elliott PM, Anastasakis A, Borger MA, Borggrefe M, Cecchi F,
A two month male baby with Alagille Syndrome, posted for pyeloplasty: Anaesthesia management

Sir,

A 2-month-old male baby weighing 4 kg was brought to the hospital with a history of fever for 2 days and jaundice for one week. He was born to non-consanguineous parents and was delivered by emergency caesarean section in view of meconium-stained liquor. The baby had physiological jaundice and respiratory distress at birth and was treated in the neonatal intensive care unit (NICU) before discharge. The child was immunised appropriately.

The relevant laboratory reports were: total leucocyte count 16900 cells/cu mm, total bilirubin 14 mg/dL (direct 5 mg/dL), serum glutamic oxaloacetic transaminase (SGOT) 649 U/L, alkaline phosphatase (ALP) 438 IU/L, activated partial thromboplastin time (aPTT) 41.7 s.

Ultrasonography (USG) of abdominal-pelvis showed gross right hydro-uretero-nephrosis with cortical thinning and mild hepato-splenomegaly. Tc ethylene dicysteine scan showed right pelvic-ureteric junction obstruction. Tc 99 mebrofenin hepatobiliary scan showed enlarged liver with impaired hepatocyte function. Mildly dilated right atrium (RA) and right ventricle (RV), moderate pulmonary branch stenosis predominantly at the left pulmonary artery (LPA) with a gradient of 40 mmHg, a small patent foramen ovale (PFO) with the left to right shunt and mild tricuspid regurgitation were seen on echocardiography.

The child was stabilised with vitamin K, fresh frozen plasma (FFP) and antibiotics. A diagnosis of Alagille syndrome with obstructive uropathy and unconjugated hyperbilirubinemia was made, and the child was scheduled for right pyeloplasty. Relevant findings on pre-operative assessment were prominent forehead and pointed chin with normal temporomandibular joint mobility.

The standard monitoring—electrocardiography (ECG), oxygen saturation (SpO2), non-invasive blood pressure (NIBP), precordial stethoscope, temperature and capnography was started in the operating room. The baseline vitals were stable. A 22- G intravenous cannula was secured in the right upper limb. 100% oxygen was given by facemask. A radiant warmer with a preset temperature of 37°C was used. The child was premedicated with 0.4 mg of ondansetron, 0.04 mg of glycopyrrolate, 0.1 mg of midazolam and 8 mcg of fentanyl intravenously.

General anaesthesia was induced with 10 mg of propofol, and 5 mg of succinylcholine was used to facilitate tracheal intubation (uncuffed size 3 endotracheal tube). As the pre-operative values of platelet count and prothrombin time—international normalised ratio (PT-INR) were normal, caudal block was given with 2.5 mL of 0.25% bupivacaine. Anaesthesia was maintained with sevoflurane in oxygen-nitrous oxide, and atracurium (2 mg initially and 0.5 mg top up) was used for muscle relaxation.