The Utility of Targeted Perioperative Transthoracic Echocardiography in Managing an Adult Patient with Anomalous Origin of the Left Coronary Artery-pulmonary Artery for Noncardiac Surgery

Abstract
Congenital coronary artery anomalies as a whole are uncommon. Abnormal origin of the left coronary artery from the pulmonary artery (ALCAPA) is probably the most common congenital coronary defect. An overwhelming majority of the patients with untreated ALCAPA do not survive to adulthood. As yet, there is no consensus on the management of adults with ALCAPA. We describe a patient with breast malignancy and incidentally detected ALCAPA; primacy was given to treatment of the oncologic condition as a first step. Anesthesia management was focused on maintaining adequate collateral coronary perfusion and avoidance of excessive loading of the left ventricle. This was achieved using a simplified transthoracic echocardiography (TTE) protocol at the time of induction of anesthesia; TTE was also used to reconfirm the absence of disturbances in myocardial function at the end of surgery. We suggest the routine use of TTE in managing perioperative care in low resource settings when the underlying cardiac disease is rare and the evidence base if often insufficient.

Keywords: Abnormal origin of the left coronary artery from the pulmonary artery, anesthesia, noncardiac surgery, transthoracic echo

Introduction
Abnormal origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare entity (1:300,000 live births). Most deaths occur in infancy and survival into adulthood is only an estimated 10%. Most patients present in early infancy with left ventricular dysfunction, mitral regurgitation, or sudden cardiac death. Adult survival is predicated by extensive collateral supply from the right coronary artery (RCA) which is then large, dilated, and tortuous while the left coronary artery (LCA) is small and thin.

Breast cancer is an ominous malignancy with any delay in diagnosis and surgery impacting survival. On the other hand, the preferred management of ALCAPA in adults is less certain. The consultation between cardiology, surgery and anaesthesia teams chose therapeutic intervention for the breast malignancy to be accorded immediate priority. We describe the successful conduct of anesthesia for ALCAPA below using a minimalist approach based on transthoracic echocardiography (TTE) which has implications on cost mitigation for low-middle-income economies with limited healthcare spend such as ours, in India.

Consent
The indexed patient has reviewed the report and provides written consent to potential publication of the report. As authors, we have removed all case identifiers from the images and the report that would be prejudicial to patient privacy. Each of us has contributed variously to this paper as described in the attestation (vide supra).

Case Report
A 54-year-old female patient was referred to our tertiary care university hospital with a history of hospital admission for unstable angina global repolarization and surgical management of a Stage IIB breast malignancy. The episode of unstable angina was managed in a hospital conservatively. Clinical details and electrocardiographic abnormalities had previously led to a diagnosis of acquired coronary stenosis. In view of urgent need for targeted echocardiography, transthoracic (TTE) was performed.

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breast cancer surgery, she chose referral to our hospital for simultaneous management of both conditions. The only current symptoms were dyspnea on climbing two flights of stairs and no orthopnea or paroxysmal nocturnal dyspnea or chest pain. Clinical examination of the cardiovascular system was remarkable for a pansystolic murmur suggestive of mitral regurgitation. The first TTE demonstrated a dilated left heart, mildly diminished global wall motion, a left ventricular ejection fraction of 40% (modified Simpson method), moderate to severe mitral regurgitation, and an estimated right ventricular systolic pressure of 45 + right atrial pressure mean. The tricuspid annular plane systolic excursion was noted to be 23 mm. The TTE also recorded that the LCA orifice was not visualized and the RCA was deemed exceptionally dilated and ecstatic raising suspicion of a diagnosis of ALCAPA. Stress perfusion with technetium-99m sestamibi showed a reversible perfusion deficit in the territory expected to be supplied by the LCA. A coronary angiogram, in fact, demonstrated the origin of the LCA from the pulmonary artery [Figures 1 and 2] and the dilated RCA. Major hematologic and biochemical screening was unremarkable, with a hemoglobin level of 11 g/dL. Consultation between cardiology, cardiac surgery, oncologic surgery, and anesthesia teams consensually agreed to accept the patient for a total mastectomy, with axillary clearance as a priority procedure. ALCAPA management was to be done as a second-stage procedure.

On the day of surgery, after standard preoperative fasting, the anesthesia team performed a focused cardiovascular ultrasound (FoCUS) in the holding area. The results of this examination concurred with the previous expert cardiologist report noting well preserved right ventricular function (tricuspid annular plane systolic excursion of 24 mm), mild left ventricular dysfunction (left ventricular end diastolic dimensions of 63 mm), fractional shortening of 25%, fractional area change of 40%, and a wall motion score index (WMSI) of 2. The left ventricular end diastolic area on the parasternal short-axis view was assessed to be 14 cm².

Intravenous access and a radial arterial catheter were secured under local infiltration anesthesia. Ultrasound-guided paravertebral block was performed at the T4 vertebral level with 0.5% ropivacaine (0.3 mL/kg) in the sitting position. Standard monitors such as 5-lead electrocardiogram (ECG), pulse oximetry, and end-tidal carbon dioxide (ETCO₂) and anesthetic gas were applied. Infective endocarditis prophylaxis was administered according to extant guidelines/anesthesia was induced with incremental doses of intravenous propofol. Intravenous Ringer lactate fluid boluses of 100 mL and 50 μg boluses of phenylephrine were used to treat >10% changes in mean arterial pressure from baseline. The decision to infuse fluid or inject vasopressor was based on repeated viewing of the left ventricle in the parasternal short axis window at the level of the papillary muscle; the left ventricular end diastolic area was attempted to be maintained within the normal limits of 14–18 cm². Any change in global or regional ventricular function on a qualitative basis was also assessed. The stroke volume obtained from an apical five-chamber view was 47 mL, and the cardiac index was 2.3. If need be, this measurement could have been repeated to optimize systemic hemodynamics. Endotracheal intubation was facilitated with 0.1 mg/kg of vecuronium bromide and thereafter titrated to 1–2 responses on a train-of-four stimulation. Anesthesia was maintained with isoflurane (end-tidal anesthetic agent of 1–1.5%) in an air-oxygen mixture (fractional concentration of oxygen 45%) and mechanical ventilation was adjusted to achieve an ETCO₂ of 35 mm Hg. An arterial blood gas sample was obtained after attainment of stable hemodynamics to establish the arterial to ETCO₂ gradient which was then determined to be 5 mm Hg; in addition, this was also used to determine the arterial oxygen tension which was 120 mm Hg. Surgery lasted for 95 min; at the end of the

![Figure 1: Dilated right coronary artery](image1)

![Figure 2: Left coronary artery filled by collaterals from the right coronary artery](image2)
procedure, neuromuscular block was duly reversed and the trachea extubated. Temperature monitoring and forced air warming were used to maintain a nasopharyngeal temperature of 36°C. In addition to routine limb lead II and precordial lead V5 visualization on surface ECG, real-time ST segment monitoring (with an alarm set at 2 mm below the patient’s baseline and measured at J + 80 ms) was also commenced and monitored throughout the procedure on a Datex-Ohmeda S/5 monitor (GE Healthcare, Madison WI, USA). Invasive right heart catheters were not placed as we surmised that owing to the low-risk nature of breast surgery with minimal fluid shifts, hemodynamic perturbations would be limited to use of anesthesia medications and positive pressure ventilation. Hemodynamic parameters were unchanged (maximum deviations of 10% from baseline in the heart rate and mean arterial pressure) throughout the procedure and in the postoperative anesthesia care unit.

An intravenous infusion of morphine at the rate of 40 µg/kg/h was commenced with the analgesia protocol prescribing in addition to 4 g of intravenous paracetamol every 24 h. Boluses of fentanyl (2/µg/kg) were used if the numerical rating score for pain exceeded 4. Echo examination was repeated in the postoperative holding area which was unremarkable and not different from the preoperative examination in terms of either wall motion or overall ventricular function. A point-of-care cardiac troponin visual estimation kit (TROP T®, Roche Diagnostics, Basel Schweiz, cutoff 100-ng/mL) was measured on postoperative days (PODs) 0–2 to detect occult myocardial damage and screen for further quantitative troponin estimation. All of these results were negative. The patient was discharged from the postanesthesia care unit at 24 h and hospital on POD 3; recovery was unremarkable and she has since been referred to a center of her choice for surgical correction of her cardiac condition.

Discussion

The global burden of adults with grown-up congenital heart disease is reaching epidemic proportions. Patients are likely diverse with either residual lesions, complications of repair, or persistent irreversible change from the preoperative state. Specific to the index case, it is uncertain as to whether translocation, coronary artery bypass graft to the LCA branches, or ligation offers the maximum benefit in adults. ALCAPA has a notoriously variable life history ranging from sudden cardiac death, presentation with cardiac failure, or mitral regurgitation at some point of time in the natural history of the disease or serendipitous discovery in asymptomatic patients. At birth, the physiologic consequences of the abnormal coronary anatomy are silent related to the high pulmonary artery pressures (PAPs); with the decline in PAP, oxygen supply in the LCA territory is dependent largely on the collateral supply from an abnormally dilated right counterpart and less so on the mixed venous oxygen tension. The collaterals also potentially present a left to right (L-R) shunt between the systemic circulation represented by the RCA and the lower resistance pulmonary circulation. Besides the consequences of the L-R shunt itself, this would also obviously precipitate myocardial ischemia in the LCA territory. While most reports consider the myocardium hypoperfused but viable, there appears a component of fibroelastosis from chronic ischemia which is a substrate for potentially fatal arrhythmia. Mitral regurgitation can result from global left ventricular dysfunction or papillary muscle ischemia-infarction. Life expectancy as a rule is shortened.

Focus on the following goals in the intraoperative period seems reasonable: ensure adequate collateral flow through maintenance of forward cardiac output and judicious use of a vasopressor, prevent coronary steal from sudden or excessive drops in the pulmonary vascular resistance by maintaining a reasonable arterial saturation without causing hyperoxia or hypocapnea (and calibrating the intraoperative monitors, viz., end-tidal capnogram and pulse oximeter to an arterial blood gas sample). Finally, it is important to prevent surges in systemic vascular resistance that could compromise forward output of an abnormal left ventricle, increase mitral regurgitation besides decreasing the net coronary artery perfusion pressure by increased left ventricular end diastolic pressure and lowered mean arterial pressure. This was achieved by ensuring adequate end-tidal anesthetic agent depth, adequate pain control with a multimodal analgesia regime in the intraoperative and immediate postoperative period, and rigorous maintenance of core temperature during the procedure. Also important is to titrate preload in a compromised ventricle, particularly during anesthesia induction, when fluid shifts with the commencement of mechanical ventilation and the action of intravenous agents are maximal. This was effectively done using transthoracic echo and is probably the first formal description of its kind.

Volatile agent anesthesia is believed to have pleiotropic effects on protecting against myocardial ischemia. Since clinical outcomes are not conclusive, the current guidance on intravenous versus volatile agent choice is equivocal. Yet, volatile agent anesthesia is more easily titrated using the end-tidal anesthetic gas value.

Echocardiography has been adapted to the perioperative period with the FoCUS approach which we used with additional emphasis on the WMSI, the end diastolic area on parasternal short axis view (as a surrogate of preload) and the fractional area change (as an estimate of contractility). In our case, it was particularly used to detect the status of left ventricular preload (as adjudged by the end diastolic area on the parasternal short-axis view at the level of papillary muscle), any changes in global or regional ventricular function, as the patient was at risk. We
also had some measure of systemic hemodynamics by a measurement of stroke volume.

Finally, we chose to do serial measurements of cardiac troponins to detect occult myocardial injury; the point of care test has a cut-off of 100/ng/mL; if positive, a quantitative level was to be sought. However, the publication of VISION (which postdated the index case) would likely have warranted quantitative assay, in any case, in view of the extremely low cutoff values above which this study has demonstrated risk.[18]

In conclusion, we describe the successful conduct of anesthesia in an adult with incidentally diagnosed ALCAPA; illustratively, we suggest that for rare conditions such as these where evidence is scarce principles based on pathophysiology, Ab Initio can be used for satisfactory guidance and outcomes. Limited TTE is a nonresource intensive, useful monitor in hemodynamic management in the perioperative period in such cases, particularly when fluid shifts are small or negligible. It can potentially be adapted to the pathophysiology of individual lesions, particularly when guidance on the optimal management of these patients during noncardiac surgery is scarce.

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Conflicts of interest

There are no conflicts of interest.

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