The anaesthetic implications of Kartagener’s syndrome are varied. The anaesthetist might be involved with patients who have sinus surgery, pulmonary surgery, infertility investigations or possibly cardiac surgery. Of primary importance will be assessment of pulmonary and cardiac structure and function, and also prevention of pulmonary complications in the bronchiectatic patient.

Physiotherapy, postural drainage, antibiotics, bronchodilators and incentive spirometry all have a role perioperatively. Where possible, local or regional anaesthesia is to be preferred to general anaesthesia. In thoracic surgery, the anatomy of the bronchi should be considered before selecting a double lumen tube.

Knowledge of the position of the abdominal organs and of the branching pattern of the main stem bronchi is important in categorizing malpositions; and these must be borne in mind. When dextrocardia occurs with situs inversus, the heart is most frequently normal. When dextrocardia occurs without situs inversus, when the visceral situs is indeterminate (situs ambiguous), or if isolated levocardia is present, associated, often complex, multiple anomalies are usually present. Dextrocardia with complete situs inversus occurs in approximately 2 per 10 000 births. The incidence of congenital heart disease is low being about 3%. Kartagener’s syndrome will occur in about 20% of patients. Thus, the incidence being approximately 1 in 50 000 births. Our patient did not have an echocardiogram and on clinical grounds was assumed to have a totally normal heart.

In contrast, dextrocardia with situs solitus or situs ambiguous is less common (1 per 20 000 births) and the incidence of congenital heart disease is extremely high, probably 90% or greater. Dextrocardia with situs solitus usually, although not invariably, associates with severe complex cardiac abnormalities. They are most commonly transposition of the great arteries, double outlet right ventricle, ventricular septal defect, single ventricle and pulmonary stenosis or artresia. In patients with dextrocardia and situs ambiguous, polysplenia or asplenia may be present in association with complex multiple cardiac abnormalities. These include a combination of systemic and pulmonary venous abnormalities, defects in the ventricular and atrial septa and endocardial cushion defects. There may be pulmonary artery obstruction and maldevelopment of the great arteries.

The incidence of isolated levocardia (a left-sided heart) with situs inversus is about 0.6% per 10 000 births and more than 90% have serious heart disease.

References


Acute right-to-left inter-atrial shunt; an important cause of profound hypoxia

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Three patients presented to our intensive care unit over a 3-yr period with profound hypoxia resulting from acute right-to-left inter-atrial shunt (RLIAS). Patient 1 was a 67-yr-old male with an atrial septal defect who became hypoxic and developed the rare sign of platypnoea following elective repair of an abdominal aortic aneurysm (breathlessness made worse when upright and relieved by lying flat). Patient 2 was a 38-yr-old female who developed platypnoea and...
hypoxia secondary to a patent foramen ovale (PFO) and pericardial effusion. Patient 3 was a 46-yr-old male with a PFO who developed hypoxia without platypnoea because of multiple pulmonary emboli following right hemicolecotomy. These case reports illustrate the need to consider RLIAS as a cause of hypoxia of sudden onset. Early use of bubble contrast echocardiography is indicated.

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Shunting of blood across a defect between the two sides of the heart is usually directed by the left-to-right pressure gradient. A large and persistent left-to-right shunt can increase pulmonary blood flow, which over months or years leads to microvascular changes, increased pulmonary vascular resistance and elevated pulmonary artery pressure. When the pulmonary arterial pressure approaches the systemic arterial pressure, the direction of the shunt reverses (the Eisenmenger syndrome). This chronic development of right-to-left shunt (RLIAS) is accompanied by electrocardiographic changes of atrial enlargement and right ventricular hypertrophy. Chest radiography reveals prominent pulmonary arteries.

In contrast, the adult patient with an asymptomatic interatrial defect may develop an acute RLIAS following a cardiac or respiratory insult. Acute RLIAS is a separate clinical entity from the Eisenmenger syndrome. Acute RLIAS is often overlooked as a cause of hypoxia but early diagnosis can direct therapy and avoid unnecessary invasive investigations. The three cases reported here illustrate the need to consider RLIAS in the differential diagnosis and investigation of profound hypoxia.

Case reports

Patient 1

A 67-yr-old man presented for elective repair of an abdominal aortic aneurysm. He had a past history of hypertension, transient ischaemic attacks and a left hemiparesis from which he had made a full recovery. His exercise tolerance was good and a preoperative multiple uptake-gated acquisition scan demonstrated a left ventricular ejection fraction of 67%. Anaesthesia and surgery were uneventful. The peroperative cardiac index was 3–4 litres min\(^{-1}\) m\(^{-2}\) and pulmonary arterial pressures ranged between 28/8 and 52/24 mm Hg. He required admission to intensive care unit (ICU) 3 days postoperatively with hypoxia (P\(_{a_o}\)=5 kPa) on room air, pulmonary oedema, blood pressure of 90/60 mm Hg and oliguria. He required tracheal intubation, positive pressure ventilation, diuretics and inotropes. His oxygenation improved, allowing tracheal extubation after 3 days but after 10 days his P\(_{a_o}\) was 5.65 kPa and P\(_{a_CO_2}\) 2.7 kPa on a F\(_{O_2}\) 0.95. He also developed platypnoea; that is he was unable to sit upright because of severe breathlessness but he was less breathless when lying flat, particularly in the left lateral decubitus position.

Initially the underlying cause of this patient’s hypoxia was not clear. Transsthoracic echocardiography (TTE) demonstrated good left ventricular function, mild mitral regurgitation and septal hypertrophy. A lung perfusion scintigraphic scan showed no evidence of pulmonary embolism. Thoracic computerized tomography revealed non-specific bilateral shadowing of the lung fields consistent with adult respiratory distress syndrome (ARDS) or infection. Despite antibiotic therapy followed by a trial of prednisolone 70 mg day\(^{-1}\) for possible ARDS, he continued to deteriorate and required reintubation and positive pressure ventilation. A pulmonary artery flotation catheter demonstrated a cardiac index of 2.1 litres min\(^{-1}\) m\(^{-2}\), mean right atrial pressure of 8 mm Hg, mean pulmonary arterial pressure of 37 mm Hg and pulmonary artery occlusion pressure of 3 mm Hg. During inspired nitric oxide therapy at 20 p.p.m. the cardiac index increased to 3.8 litres min\(^{-1}\) m\(^{-2}\), the mean right atrial pressure was 4 mm Hg, mean pulmonary arterial pressure 34 mm Hg and pulmonary artery occlusion pressure 5 mm Hg. The P\(_{a_o}\) increased from 10.5 to 24.5 kPa on a F\(_{O_2}\) 1.0.

A cardiology opinion was sought concerning the possibility of an inter-atrial shunt, but this diagnosis was thought to be excluded by the absence of a persistent right-to-left pressure gradient; right atrial pressure range of 4–8 mm Hg with a pulmonary artery occlusion pressure range of 3–5 mm Hg. However, an inter-atrial defect was discovered fortuitously during pulmonary angiography when a catheter passed from the right atrium into the left atrium. Bubble contrast transoesophageal echocardiogram (TOE) demonstrated RLIAS with severe impairment of right ventricular function (Fig. 1).

The patient became septicaemic and died on the 26th postoperative day. Post-mortem examination revealed a secundum ASD, absent superior vena cava and anomalous venous drainage via a dilated coronary sinus. The right ventricle appeared normal and the lungs were oedematous. On histological examination, there was mild pulmonary fibrosis but no features of ARDS, organizing pneumonia, or pulmonary hypertension.
function was good. Pericardial tap brought immediate relief of the patient’s hypoxia; the \( P_{A0} \), rising from 5.8 to 30.4 kPa, on oxygen 10 litres min\(^{-1}\) by facemask.

**Patient 3**

A 46-yr-old man presented with breathlessness 4 days after right hemicolecction for caecal carcinoma. Axillary temperature was 38.5°C, he had a sinus tachycardia 135 beat min\(^{-1}\), arterial pressure 90/60 mm Hg and a respiratory rate of 32 breath min\(^{-1}\). \( P_{A0} \), was 4.1 kPa and \( P_{ACO2} \), 4.2 kPa on room air. Oxygen at maximum flow via a facemask did not improve his hypoxia and he was transferred to ICU. \( P_{FIO2} \), 0.9 with CPAP 5 cm H\(_2\)O produced a \( P_{A0} \), 7.5 kPa. The patient was not distressed despite his hypoxia and was able to tolerate lying flat for a pulmonary angiogram. Pulmonary angiogram revealed a large embolus in the left-upper lobe pulmonary artery with smaller emboli in the right middle lobe pulmonary artery and right pulmonary vessels peripherally. The catheter passed from the right atrium to the left atrium. Mean right atrial pressure was 13 mm Hg, left atrial pressure 11 mm Hg and mean pulmonary arterial pressure 27 mm Hg. Left pulmonary venous \( P_{O2} \), was 32.3 kPa but the radial arterial \( P_{A0} \), was 6.24 kPa. A Greenfield inferior vena cava filter was deployed with its base at the level of the L3-L4 disc space. A bolus of tissue plasminogen activator 5 mg i.v. plus an infusion of 5 mg h\(^{-1}\) was instituted. The patient’s oxygenation improved over the next 3 days and thereafter his recovery was uneventful.

**Discussion**

Acute RLIAS is a rare but important cause of profound hypoxia. The pathophysiology of acute RLIAS arises from an inter-atrial defect coupled with a secondary cardiac or pulmonary insult. Patent foramen ovale, which occurs in approximately 30% of the adult population,\(^4\) is present in the majority of reported cases of acute RLIAS. ASD is present in the remainder. A rise in right atrial pressure above left atrial pressure may precipitate RLIAS. This can be the result of obstruction of pulmonary blood flow by pulmonary embolus\(^6\) (Patient 3), increased transmural pressures in asthma,\(^2\) right ventricular hypokinesis following right ventricular infarction\(^6\) or coronary artery bypass grafting,\(^7\) or as a result of elevated \( P_{ACO2} \), in hemidiaphragmatic paresis.\(^5\) However, in similar cases right heart pressures can be normal, which indicates that other factors can induce RLIAS.\(^6\)\(^9\)\(^10\) Mechanical distortion of the heart can alter the relative positions of the atrial septum and the inferior vena cava (IVC) which then directs the flow of blood from the IVC towards the inter-atrial defect and into the left atrium. Mediastinal shift following lung resection is the most commonly reported cause of RLIAS.\(^11\) Other causes of distortion of cardiac anatomy with no apparent right-to-left pressure gradient include thoracic trauma,\(^12\) rupture of the papillary muscles,\(^13\) after tuberculosis,\(^14\) thoracic aortic...
aneurysm, pericardial effusion (Patient 2), and metastatic cardiac disease. The mechanism of RLIAS is unclear in cases of morbid obesity, cystic fibrosis, adult respiratory distress syndrome and exposure to low atmospheric pressure.

Platypnoea is a sign that is strongly associated with RLIAS. This sign, first described in 1949 and given the name platypnoea in 1969, occurs in approximately one quarter of reported cases of acute RLIAS. A corresponding postural change in oxygenation is known as orthodeoxia.

RLIAS is often diagnosed unexpectedly during investigation for other causes of acute hypoxia. A pulmonary angiography catheter may pass through an inter-atrial defect, as occurred in our patients, but the angiogram may be reported as normal despite the presence of a significant RLIAS. Lung perfusion scintigraphy indicates RLIAS when early activity is detected in the brain, myocardium and kidneys. It has been advocated as the investigation of choice but failed to detect RLIAS in our first and second patients and in several other reported cases. Injection of radiolabelled microaggregates into the arm has resulted in a normal perfusion scan whereas injection into the foot demonstrated shunt because of preferential flow from the IVC across the interatrial defect.

Bubble contrast echocardiography has the greatest sensitivity for detecting RLIAS and has the advantage that it is a relatively non-invasive technique that can be performed at the bedside. A syringe filled with 9 ml saline and 1 ml air is agitated, macroscopic bubbles expelled and the remaining microbubble emulsion injected i.v. The test is positive if microbubbles are seen in the left atrium (Fig. 1) within two to three cycles of the initial appearance in the right atrium.

Bubble contrast echocardiography has detected RLIAS as a first line investigation and when preceding pulmonary angiogram and lung perfusion scintigram have both been falsely negative. Injection of contrast via the leg may increase the sensitivity of the test as most shunting occurs via the IVC. RLIAS is exacerbated when intrathoracic pressure is increased by PEEP in the artificially ventilated patient and during coughing or the Valsalva manoeuvre. Each of these manoeuvres can increase the likelihood of detecting RLIAS by echocardiography.

Bubble contrast echocardiography is a safe technique widely reported in the investigation of chronic RLIAS, particularly in patients who are thought to have developed paradoxical embolism. Paradoxical embolism is not commonly associated with profound hypoxia as these patients present with systemic embolic phenomena. Patient 1 had a past history of cerebrovascular events and whether these were caused by paradoxical emboli is an interesting matter for conjecture.

Treatment of RLIAS is treatment of the underlying cause and/or closure of the PFO or ASD. Successful closure of the inter-atrial defect is frequently reported although some patients have died following its closure because of right heart failure. Temporary closure may be achieved with a balloon catheter but definitive closure requires either open surgery or a percutaneous technique using an occlusive device.

Patient 1 developed acute RLIAS and platypnoea. The nature of the precipitating cause remains uncertain. However, the raised perioperative pulmonary arterial pressures and the subsequent beneficial effect of nitric oxide suggests that pulmonary vasoconstriction was a contributing factor. The diagnosis of RLIAS was discounted because of the absence of a significant right-to-left pressure gradient between the atria. By the time RLIAS was diagnosed, the patient’s condition precluded surgical closure of his ASD. The use of bubble contrast during the patient’s first echocardiogram might have revealed the RLIAS early enough for surgery to be feasible.

Patient 2 presented with platypnoea; as this is highly indicative of RLIAS, early use of bubble contrast echocardiography was indicated. A lung perfusion scintogram failed to detect it and was falsely positive for pulmonary embolism. RLIAS was discovered during pulmonary angiography when the catheter passed through a PFO. Early use of bubble contrast echocardiography might have saved the patient from these more invasive investigations.

Patient 3 demonstrates that acute RLIAS can occur without platypnoea and without high pulmonary arterial pressures.

In conclusion, the diagnosis of acute RLIAS should be considered in any patient with unexplained acute hypoxia particularly if platypnoea or orthodeoxia is present. Neither pulmonary hypertension nor a pressure gradient between the right and left atria need be present. Bubble contrast echocardiography is more sensitive in detecting RLIAS than more invasive techniques and should be performed early.

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