

Intrapulmonary shunting and hepatopulmonary syndrome

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INTRODUCTION

In patients with cirrhosis who develop hypoxaemia, hepatopulmonary syndrome (HPS) must be considered in the differential diagnosis. The condition is characterized by hepatic dysfunction and/or portal hypertension, hypoxaemia and intrapulmonary vascular dilatations (IPVD). The authors report an atypical case where the diagnosis was revealed by contrast echocardiography that demonstrated marked intrapulmonary shunting.

DISCUSSION

The prevalence of HPS varies between 4% and 32% in cirrhosis, with a 1-year survival between 16–48% once the partial pressure of oxygen in arterial blood (PaO_2) is <6.67 kPa (Naeije, 2003). Progressive exertional dyspnoea and platypnoea (dyspnoea that improves when supine) are typical symptoms. Characteristically, orthodeoxia (hypoxaemia worse when upright) occurs as a result of gravity-dependent pooling of blood in the IPVD, which predominate at the lung

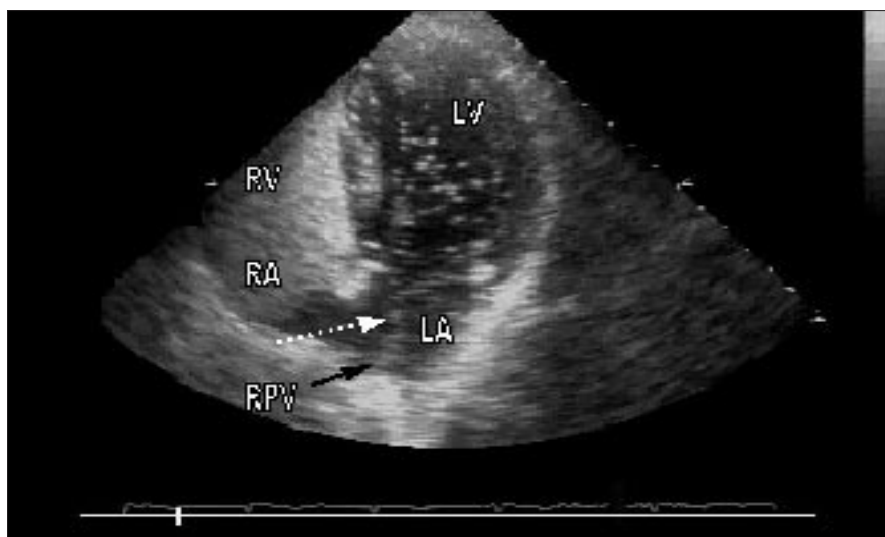


Figure 1. 2-D echocardiogram (4-chamber view) using agitated saline air bubble contrast injected into an antecubital fossa vein. There is dense opacification of the right heart and a noticeable stream of contrast microcavitations (dashed arrow) from the right upper lobe pulmonary vein (bold arrow) through the left atrium dispersing into the left ventricle.

bases, and this leads to increased intrapulmonary shunting.

The shunt is demonstrated by transthoracic contrast echocardiography, which confirms delayed visualization of agitated saline air bubbles in

the left heart chambers, 3–6 heartbeats after initial appearances in the right heart (Naeije, 2003). In patients with no shunt, the pulmonary vasculature absorbs the air bubbles, whereas with an intracardiac shunt, contrast is seen immediately in the left atrium following intravenous injection.

Atypically in this patient, contrast appeared from the right upper lobe pulmonary vein rather than the basal pulmonary veins, which would explain the absence of characteristic orthodeoxia. Carbon monoxide diffusing capacity may not always be reduced, whereas an age-corrected alveolar-arterial oxygen gradient ($AaPaO_2$) >2.70 kPa shows greater diagnostic accuracy in the assessment of HPS (Lima et al, 2004).

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CASE REPORT

A 57-year-old woman was referred for assessment of progressive dyspnoea, decreasing exercise tolerance and worsening bluish facial discoloration. She had congenital cirrhosis complicated by portal hypertension aged 4 years, for which she underwent a portacaval shunt. She was an ex-smoker of 30 years. On examination, she was centrally cyanosed and clubbed. Her jugular venous pressure was not elevated, cardiac examination was normal and there were no focal chest signs. Spider naevi were noted, but no palpable organomegaly or ascites. Her oxygen saturation, however, was 88% breathing room air, which did not alter when recumbent or supine.

A chest radiograph showed normal lung fields and vasculature, and no effusions. Renal and liver profile blood tests and international normalized ratio were normal. Arterial blood gas analysis revealed: pH 7.43 (normal 7.35–7.45), PaO_2 7.21 kPa (normal >10.6 kPa), $PaCO_2$ 3.96 kPa (normal 4.6–6.0 kPa) breathing room air. Her $AaPaO_2$ was 7.8 kPa (predicted age-corrected $AaPaO_2$ 2.10 kPa). PaO_2 increased to 69.8 kPa breathing 100% inspired oxygen delivered using continuous positive airway pressure. Her calculated intrapulmonary shunt fraction was 7% (normal $<5\%$). Pulmonary function tests showed normal spirometry and preserved lung volumes, however, carbon monoxide diffusing capacity was only 38% predicted, adjusted for haemoglobin 17.8 g/dl.

Transthoracic echocardiography demonstrated normal structure and function of all cardiac chambers. Contrast echocardiography established the presence of intrapulmonary shunting (Figure 1) and confirmed the diagnosis of hepatopulmonary syndrome. She was established on long-term oxygen therapy and is currently under consideration for a liver transplant.

An inadequate response to breathing 100% inspired oxygen, $PaO_2 < 40$ kPa, suggests the presence of an anatomical shunt (Lima et al, 2004). This patient demonstrated a good response and had a marginally elevated calculated shunt fraction (Gossage and Kani, 1998). This implied IPVD (physiological shunt) rather than arteriovenous (AV) communications (true anatomical shunt) underlying the cause of her intrapulmonary shunt. In the latter, pulmonary angiography may be useful as discrete AV communications may be amenable to embolization (Hoepfer et al, 2004). A computed tomographic pulmonary angiogram undertaken in this patient did not reveal AV communications.

Radionuclide lung perfusion imaging with technetium-labelled macroaggregated albumin can also quantitate the intrapulmonary shunt fraction, which has prognostic implications, as mortality in HPS is associated with higher shunt fractions (Arguedas et al, 2003).

Currently, there is no effective pharmacological treatment of HPS (Hoepfer et al, 2004). Orthotopic liver transplantation is the only definitive treatment option with good resolution of IPVD, although it may take several months for reversal of abnormal oxygenation (Krowka et al, 2004). Perioperative mortality, however, remains high, particularly because of advanced hypoxaemia. Early diagnosis, therefore, remains

essential and physicians should have a high index of suspicion. **HM**

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IN THE PUBLIC'S VIEW...

What is health?

Panorama (BBC1, 20 February) gave us 'Life in an NHS hospital'. It wasn't any old hospital but the three star North Bedford NHS Trust. North Bedford is remembered for the 'bodies in the chapel' episode. Panorama described this as the hospital's 'public disgrace', which led to its being awarded no stars and its then chief executive resigning. But it wasn't quite like that. The mortuary was full, and they had to do something with the bodies. A chapel seems to me a reasonably respectful place to put bodies. The real scandal was the staff who leaked the story to the press, and the people who photographed the purposely disarrayed bodies. It struck me at the time that a number of people should have received their P45s, and the media should have been more realistic and more sensitive. As that is not what the media are interested in, what we got was public disgrace for the trust.

However, from that, North Bedford gained a new chief executive, Andrew Reed, and a lot of attention from the Department of Health. Their prize was three stars, awarded for meeting government targets. Reed was clear that targets were important and had had an effect. He had stressed to the staff that they were meeting targets not for the

government, but for the better care of the patients. To that extent, targets can appear a good thing, but the programme slowly diluted the message by showing the perverse incentives and sometimes plainly idiotic consequences of having targets that have to be met.

Cutting deaths from cardiac disease is high on the government's list. So, equipped with a new catheter laboratory, and having to comply with maximum waits for exercise testing, North Bedford gives a timely test to everybody with chest pain. They do not have enough trained staff to interpret the tests, so some of the tests are pointless, but the only thing that matters is whether the test has been done.

The 2-week cancer wait works well for those whose symptoms suggest cancer (though whether it improves survival is doubtful) but, as the colorectal surgeon said, many of those having suggestive symptoms do not have cancer, and some of those with not quite suggestive symptoms do. As his waiting list is not under his direct control any more, but generated by non-medical waiting list coordinators, patients about whom he would be suspicious are sent routine appointments.

At North Bedford, as with many hospitals, imaging is a bottleneck. They got

money for an magnetic resonance imaging (MRI) scanner, but it is in a van in the car park. There is no money for infrastructure, it is not nice for the patients, and radiologists are leaving because the working conditions are poor.

But, in general, because of the government's largesse, North Bedford is better than it was and the staff are dedicated and keen. They have worked hard, more patients are being seen, and more operations are being done. Result: financial shortfall, and the reward for all the hard work is likely to be loss of a star. What a morale booster that will be.

As John Appleby of the King's Fund, one of two independent health analysts interviewed for the programme, said, 'Has Labour's money helped? Well, we don't really know, because we don't know what health is. We're measuring waiting times, but we are not actually measuring health.'

Half my operating lists so far this year have been curtailed because there are medical patients in surgical beds. I do not know what that means for our population's health, but I do know what it does to targets. **HM**

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