

Case Reports

A case of orthodeoxia platypnoea in a patient with adult polycystic kidney and liver disease with a patent foramen ovale

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Key Learning Points

1. Orthodeoxia Platypnoea describes the phenomenon whereby desaturation and dyspnoea occur in the upright position in association with an intra atrial shunt.
2. Closure of the intracardiac defect is recommended to improve symptoms and prevent development of ischaemic strokes secondary to paradoxical embolism.
3. A history of postural change in symptoms should be sought for all patients presenting with breathlessness where the diagnosis is unclear.

Abstract

We describe a case of a patient with polycystic kidney and liver disease presenting with a 4 year history of shortness of breath. This was caused by a liver cyst pressing on the inferior vena cava, right atrium and ventricle, leading to a right to left shunt across a patent foramen ovale. This is consistent with the condition Orthodeoxia Platypnoea, which occurs when desaturation and dyspnoea occur in the upright position in association with an intra atrial shunt.

Case Report

A 66-year-old female patient presented on the acute medical take with worsening exertional dyspnoea. Her exercise tolerance was limited to 20 yards. She had been investigated for her increasing dyspnoea in the preceding 4 years; investigations had included a normal coronary angiogram and a Trans Thoracic Echo (TTE) showing good LV function, mildly dilated left atrium (4.7 cms), mild mitral regurgitation and good right ventricular function with a normal sized right heart. The dyspnoea had been attributed to her obesity in the absence of any other explanation.

Her past medical history included adult polycystic kidney disease, bilateral oophorectomies for benign cysts, hypertension and a recent Transient Ischaemic Attack. She had never smoked and had no previous respiratory disease. Her only risk factor for thromboembolic disease was her obesity. Clinical examination was unremarkable.

At the time of admission she had oxygen saturations of 86% on air and 93% on 10 litres of oxygen. Arterial blood gases on air showed a pH 7.46 kPa, PO₂ 5.22 kPa, PCO₂ 3.71 kPa. Her spirometry demonstrated FEV1/FVC 1.95 litres/2.25 litres (87%) with no bronchodilator reversibility (predicted FEV1 = 2.23). No abnormality was seen on her chest X-ray.

She underwent CT Pulmonary Angiography, which showed no pulmonary emboli and normal lung parenchyma. Note was made of large hepatic and renal cysts and that the IVC was compressed by the liver cyst (Figure 1).

Her dyspnoea remained unexplained and the possibility of an intracardiac shunt was considered. Cardiological assessment took place and further investigations arranged. A repeat TTE was unchanged from previous appearances. Trans Oesophageal Echo and cardiac catheterisation were performed, showing normal right-sided pressures and saturations but that left sided saturations were reduced to a PO₂ of 8.8 kPa. Note was also made of a large cyst compressing the right atrium and ventricle but with no evidence of right atrial or ventricular collapse. No shunt was visualised.

She was discharged home on oxygen and a second opinion was sought from a tertiary referral centre. Shortly before her outpatient review she volunteered

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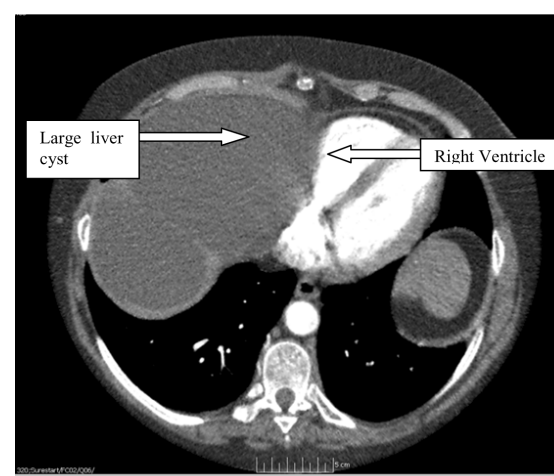


Figure 1. CTPA showing large liver cyst compressing right heart.

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some extra history to her general practitioner, stating that all the tests had been carried out whilst she was lying down, but that her symptoms only occurred when upright.

The diagnosis of 'Orthodeoxia Platypnoea' was made and confirmed by a contrast ECHO showing a liver cyst compressing the right atrium and reducing IVC inflow across a large patent foramen ovale. It was noted that she became profoundly cyanotic on standing.

Her patent foramen ovale was closed surgically, and was noted to be 12mm in diameter.

Following surgery her exercise tolerance had significantly improved, she no longer required domiciliary oxygen. Resting oxygen saturations on air were 96%. She felt the best she had done in the past 4 years.

Discussion

Orthodeoxia platypnoea is an uncommon condition where dyspnoea and hypoxia are worse in the upright position. It can prove difficult to diagnose, particularly when the association with posture is not initially reported. Traditionally investigations undertaken in hospital tend to be performed with the patient seated or supine.

In order to occur there needs to be an intra-cardiac shunt which becomes apparent in the upright position by the physical affects of an additional mechanism. This phenomenon is most commonly described in patients with a history or pulmonary disorders, such as recurrent pulmonary embolism,¹ emphysema,¹ postpneumonectomy,¹ ARDS,¹ hepatopulmonary syndrome,¹ pulmonary AV malformations,¹ pneumocystis pneumonia² and Idiopathic Pulmonary Fibrosis.³

It has also been described without associated pulmonary disease in hydatid liver cyst,⁴ aortic root dilatation,⁵ aortic aneurysm⁶ and pericardial effusion.⁷ As in our case, pulmonary embolism is often initially suspected.⁸

This case represents the first report of the combination of a large liver cyst caused by PCKD and a co-existent PFO leading to the development of Orthodeoxia Platypnoea. After surgical repair of the PFO there was a good recovery.

It is not clear why there is such marked positional variation in the right to left shunt. One postulated mechanism is that in the upright position the right atrium is distorted by pressure from the liver cyst causing a directed flow of blood towards the patent foreamen ovale. It is also possible that the liver cyst caused a change in the anatomical relationship of the inferior vena cava to the patent foreamen ovale, once again leading to directed flow from the right side of the heart to the left.⁹

Studies have recommended closure of the intracardiac defect to help resolve the orthodeoxia platypnoea and prevent development of ischaemic strokes secondary to paradoxical embolism.^{1,8} Our patient had already experienced one transient ischaemic attack prior to diagnosis and subsequent closure of her patent foreamen ovale.

We believe this case illustrates how important it is to explore the history carefully, asking specifically about postural changes in patients with unexplained breathlessness. It also demonstrates that clinicians should be prepared to question negative test results which do not fit with the clinical scenario: an intracardiac shunt had been suspected, but was discounted on the basis of a normal echocardiogram.

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