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CASE STUDY

Cardiac cause of hypoxaemia in a kyphoscoliotic patient

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Cardiac cause of hypoxaemia in a kyphoscoliotic patient . I. Herry, B. Iung, J.Y. Piechaud, F. Saidi, C. Mayaud, A. Boissonnas. ©ERS Journals Ltd 1999.

ABSTRACT: This report presents the case of a 73 yr-old female in whom kyphoscoliosis, osteoporotic vertebra compression fractures and sternal injury resulted in severe respiratory failure and hypoxaemia. Pulmonary function testing showed moderate restrictive pattern and rare mismatches were found on lung ventilation/perfusion scanning. Transoesophageal echocardiography with contrast studies showed abnormal anatomic mediastinal interactions which led to right-to-left interatrial shunt, through patent foremen ovale. First-intention treatment, because of orthopaedic and respiratory surgical restraints, was to close the shunt using transcatheter devices. Follow-up after 6 months demonstrated that these interauricular umbrella devices corrected arterial hypoxaemia. True right-to-left interatrial shunts can be found in kyphoscoliotic patients, as a result of thoracic deformation, and can be safely treated with percutaneous trancatheter closure.
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In most adult cases, patent foramen ovale (PFO) with right-to-left interatrial shunt (RLIAS) is associated with an elevation of right-sided cardiac pressures, such as pulmonary embolization, pulmonary hypertension or pulmonary valvular stenosis, chronic obstructive disease, or congestive heart failure. Few reports describe PFO and RLIAS in adults in spite of normal pulmonary pressures, mostly following pneumonectomy [1].

In only one previously reported case, PFO was associated with an elongated aorta pressing down on the right atrium after vertebral compression fractures [2], resulting in RLIAS. In the present case, osteoporotic size shortening and recent sternum fracture may have caused changes in mediastinal interactions and foramen ovale reopening. Percutaneous transcatheter closure was chosen and successfully corrected hypoxaemia.

Case report

A 73 yr-old female was hospitalized in February 1998 for worsening dyspnoea and syncope. She had never smoked, had no previous medical history and had never presented with dyspnoea before December 1997. She had a painful sternum fracture related to a car accident in December 1997, concomitant with several osteoporotic compression fractures. After the car accident, the patient experienced pain and felt moderately dyspnoeic, with the dyspnoea rapidly worsening. At the time of hospitalization, clinical examination disclosed a short kyphoscoliotic patient, breath rate was 40 breaths·min^{-1}, pulse rate 93 beats·min^{-1} and blood pressure 130/80 mmHg. Sternal pain had abated. Temperature was 37°C and cardiac and pulmonary auscultations were normal. Clinical examination did not show any sign of pulmonary hypertension or right heart failure.

Chest radiograph disclosed clear lung parenchyma, normal heart size and kyphoscoliotic thorax. Kyphosis was predominant, with an 85° Cobb angle (a measure of thoracic spinal deformity [3]), and the sternum was consolidated. Profile disclosed horizontalization of the descending thoracic aorta, behind the cardiac silhouette. Arterial blood gases breathing room air showed pH 7.44, arterial oxygen tension (P_{a,O_2}) 5.7 kPa (43 mmHg), arterial carbon dioxide tension (P_{a,CO_2}) 2.8 kPa (21 mmHg) and arterial oxygen saturation (S_{a,O_2}) 84%.

Angioscan was normal, but the descending aorta was distorted and horizontalized behind the right atrium. Veinous ultrasonography of the legs was normal, perfusion/ventilation scan was considered as a low probability for thromboembolic disease. Pulmonary function testing disclosed a restrictive pattern, pulmonary total capacity was 67% of predicted (table 1). Arterial blood specimen breathing 15 L·min^{-1} oxygen showed P_{a,O_2} 8.8 kPa (66 mmHg) and P_{a,CO_2} 4.5 kPa (34 mmHg).

Transoesophageal echocardiogram (TOE) was performed, with contrast studies, which showed a patent foramen ovale at the anterior part of the right atrium. The aorta was pressed down on the right atrium, the atrial septum was deformed under pressure and blood was pushed into the left atrium. Left ventricular function was normal, the right ventricle was not dilated, pulmonary valve was normal and systolic pulmonary artery pressure was 40 mmHg. Echocardiographic and catheter studies confirmed the diagnosis of a 1 cm^2 off-centered PFO. PFO reopening was probably related to recent changes in mediastinal interactions after sternal and osteoporotic vertebral compression fractures.

Table 1. – Pulmonary function data of a 73-yr-old female patients presenting with worsening dyspnoea and syncope

Parameter	Patient's values	Predicted values	% predicted
TLC L	2.88	4.31	67
RV L	1.65	1.82	91
FRC L	1.98	2.50	79
FVC L	1.22	1.70	71
FEV_1 L	0.89	1.36	66
FEV_1/FVC %	73	80	

TLC: total lung capacity; RV: residual volume; FRC: functional residual capacity; FVC: forced vital capacity; FEV_1: forced expiratory volume in one second. The armspan of 1.48 m was used for calculating the predicted values of respiratory function [4].

Because of the patient's age and deformation, transcatheter occlusion of this PFO was chosen. Two umbrella-shaped devices were needed, and capillary blood oxygen saturation reached 95%. She was discharged with aspirin treatment (Aspegic° 250 mg·day^{-1}), and specific treatment for osteoporosis. Six months later, the patient was able to live at home. Arterial blood gases breathing room air disclosed P_{a,O_2} 9.2 kPa (69 mmHg), P_{a,CO_2} 4.8 kPa (36 mmHg), pH=7.40 and S_{a,O_2} 94%. Walk tests showed blood saturation at 88%, and a small residual leak persisted in front of the devices.

Discussion

Respiratory failure in kyphoscoliotic patients is multifactorial. Deformity, and poor compliance results in restrictive pattern upon pulmonary function testings and is associated with ventilation-perfusion mismatches, often pulmonary hypertension, and impaired inspiratory muscle function [5–7]. However, as demonstrated is this case and in a previous patient, true RLIAS may occur, related to mediastinal anatomic changes and interactions. Appropriate investigations, such as echocardiography with contrast studies must be performed when severity of dyspnoea and gazometric findings do not correlate with a moderate restrictive pattern. Cases of RLIAS may be discovered in this setting.

In the current patient with underlying respiratory failure, ventilatory restraints, age, osteoporosis and deformity, surgical closure of PFO is questionable. For this reason, transcatheter occlusion was chosen as a first step treatment, knowing that surgical closure of the septal foramen could be performed if transcatheter occlusion failed. This procedure is safe, as demonstrated in previous studies, and can be performed with Umbrella-Shaped-Clamshell or Buttoned devices [8–11]. In the current case, occlusion of this off-centered PFO, with small anterior atrial septal wall was difficult. First attempt to place a first umbrella-shaped device (J.Y. Piechaud) resulted in a partial correction of hypoxaemia. Because of a residual leak, a second device was then placed in front of the first, arterial haemoglobin oxygen saturation reached 95% during the procedure, and remained at 94% in the following days. Progressive endothelialization of devices might have eventually resulted in a complete disappearance of the residual shunt.

This case emphasizes the fact that patent foramen ovale may be an additional cause of hypoxaemia in kyphoscoliotic patients. In this setting, when surgery seems difficult to perform, transcatheter occlusion (even when achieved with two devices) is a safe procedure to close right-to-left interatrial shunts, although its efficacy needs to be tested in further studies.

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