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Urgent Percutaneous Closure of Patent Foramen Ovale in a Patient with Platypnea - Orthodeoxia Syndrome after Right Upper Lobectomy

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arrhythmia, which can be resolved with coronary artery bypass procedures, for example. When ischemia is not detected or resolved, other methods such as pacing suppression or balloon pump counterpulsation are considered.

(1) In a retrospective series of 21 patients, Fotopoulos et al. reported the stabilization of refractory ventricular arrhythmia using intra-aortic balloon counterpulsation, and included 3 patients without coronary artery disease, while Goyal et al. described the effectiveness of intra-aortic balloon pump for the treatment of complex refractory ventricular arrhythmia in a patient with dilated cardiomyopathy and normal coronary arteries. (1, 2)

The effectiveness of balloon pump for refractory arrhythmias in coronary patients is evident when correcting imbalance between oxygen supply and demand, due to increased coronary perfusion and reduced cardiac work. The effectiveness of the device is not so evident in non-coronary patients.

Fotopoulos, mentioned above, proposed an indirect mechanism mediated by reduction of adrenergic tone, reducing myocardial vulnerability to arrhythmias. The balloon also reduces ventricular systolic pressure, lowering wall and myocardial oxygen tension. (1) Another proposed mechanism is the direct effect of the balloon based on the concept of mechano-electric feedback or contraction-excitation feedback.

This concept is supported by studies on animals showing which post-load increases result in increased-ventricular ectopy and tachycardia.

From a physiological viewpoint, increasing myocardial stretch results in shortening of action potential duration, abnormal refractoriness and increased diastolic depolarization, while the effect of the balloon -reducing stretch and distention- benefits myocardial responsiveness.

A similar phenomenon was described in humans by Taggart et al, when detecting the proarrhythmic effect of increased loading conditions.

(1, 3) On the other hand, the use of a balloon together with complex circulatory support devices is an uncommon combination, with limited references in the literature.

Swinney et al. analyzed the use of balloon pump counterpulsation in combination with the HeartMate II continuous-flow device in 51 patients, and observed that the first one augmented systolic flow and decreased diastolic flow of the second one, by increasing pulsatility (targeted by an increased pulsatility index in the continuous-flow device console).

(4) Ma et al. analyzed the combination of extracorporeal membrane oxygenation (ECMO) with intra-aortic balloon pump in 34 patients, 15 of whom underwent ECMO first. This action involved the opening of the aortic valve, which improved with reduction of afterload due to balloon pump counterpulsation. While the devices in both references differ from the one we have described, the basic functional physiological mechanisms (continuous-flow devices) are equivalent, the opposite effect to increased afterload and its potential antiarrhythmic action resulting attractive.

(5) Hu et al. described the case of two patients with cardiogenic shock and electrical storm.

Those patients required circulatory support with ECMO, developing refractory ventricular arrhythmia which was resolved with balloon implant, as in our case. The authors posed the efficacy of the device in reducing left ventricular congestion by modifying ECMO non-pulsatile flow through the balloon.

(6) While evidences are somewhat speculative, it is evident that the use of the balloon in our patient resolved the ventricular arrhythmia in two instances, which was refractory to conventional treatment approaches. The usefulness of the balloon as antiarrhythmic treatment was evident in this complex scenario.

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Urgent Percutaneous Closure of Patent Foramen Oval in a Patient with Platypnea - Orthodeoxia Syndrome after Right Upper Lobectomy

The platypnea-orthodeoxia syndrome (POS) is characterized by dyspnea and hypoxemia in upright position, relieved by recumbence. (1) Pre-existing intracardiac shunting such as patent foramen ovale (PFO) is one of its most common causes.

Platypnea-orthodeoxia syndrome is caused by a
right-to-left shunt. (2) This shunt can occur without increased pulmonary pressures, and is due to anatomic conditions that redirect flow from the right atrium (RA) to the left atrium (LA), more evident in upright posture, although atrial pressures are higher. (1)

We present a case of POS secondary to PFO in a patient who recently underwent right lobectomy due to lung neoplasm.

His clinical presentation included rapidly progressive respiratory failure refractory to oxygen therapy, so there was an urgent need for PFO closure. The procedure was successfully performed with immediate improvement of dyspnea and hypoxemia.

A 70-year-old male patient came to the emergency room with an episode of sudden dyspnea in the upright posture. Physical examination revealed oxygen desaturation increasing with upright position to 80%, and hypoventilation in the right lung, with no signs of cardiac failure. He referred several episodes of dyspnea in upright position for a month. He had a history of hypertension, stroke of unclear etiology in 2009 (echo Doppler of supra-aortic trunks, NAD), colon adenocarcinoma treated with surgery and chemotherapy in 2009, and squamous cell carcinoma of the lung treated with right upper lobectomy 5 months ago.

On admission, a CT scan of the lungs showed volume loss of the right upper lobe, residual middle lobe collapse, and right hemidiaphragmatic elevation related to a past surgery.

No signs of pulmonary thromboembolism were observed.

Blood tests revealed polycythemia (hematocrit 41.5%), which was not present in previous tests.

In the absence of a pulmonary condition to justify the clinical manifestations and the refractoriness of hypoxemia to oxygen therapy, a transthoracic echocardiography (TTE) was performed to rule out intracardiac shunting.

Baseline TTE showed no signs of pulmonary hypertension. An agitated saline bubble test showed a massive passage of bubbles at baseline in the first and second beats, suggestive of right-to-left shunt. With these findings suggestive of POS due to PFO, a transesophageal echocardiography (TEE) was planned for PFO closure.

Due to rapidly progressive clinical deterioration in 24 hours, with severe refractory hypoxemia (65% O2 Sat) both in upright and recumbent positions, the patient was transferred to the intensive care unit under high-flow noninvasive mechanical ventilation, and an urgent TEE was performed.

Transesophageal echocardiography showed atrial septal aneurysm (ASA) with significant detachment of the oval fossa membrane and massive passage of bubbles with agitated saline from RA to LA. Given the patient’s abrupt worsening with sustained hypoxemia that could not be relieved with posture or oxygen therapy, urgent percutaneous closure of the PFO was decided. The procedure was performed under intra-}

**Fig. 1.** Chest X-ray showing volume loss of the right upper lobe, residual middle lobe collapse, and right hemidiaphragmatic elevation.

**Fig. 2.** A. Transthoracic echocardiography after bubble infusion showing massive passage of bubbles in the first beat. B. ICUS prior to PFO closure, showing patent foramen ovale (PFO) and aneurysmal interatrial septum.

Cardiac ultrasound (ICUS) and fluoroscopy.

A Figulla® Flex II 31/35 device was placed without incidence.

After PFO closure, immediate increase of O2 Sat from 66-70% to 95-97% was evidenced, with disappearance of dyspnea. Two months later, the patient remained asymptomatic, without evidence of residual shunt in TTE and with 97% O2 Sat both in upright and recumbent positions.

Platypnea-orthodeoxia syndrome can be explained by three main mechanisms: intracardiac shunting, pulmonary vascular shunting, and ventilation-perfusion mismatch.

Patent foramen ovale is the most common cause of intracardiac shunting due to its high prevalence in the general population (10-24%). (2) Hypoxemia results from significant right to left shunting.

There are certain situations that can course with flow inversion in the context of normal pulmonary pressures, due to a transient increase in right atrial
pressure (hemodynamic cause) or to a redirection of flow (anatomic distortion). (3) The extrinsic compression in the RA either by a hydrothorax or a pneumonectomy could increase intracavitary pressure, facilitating right to left shunting.

(1) On the other hand, anatomic conditions that modify the relation between the inferior vena cava (IVC) and ASA in upright position can redirect blood flow to the PFO.

(4) Several mechanisms can lead to this anatomic distortion, such as a prominent Eustachian valve, aortic dilation, or a pneumonectomy. (5) In the case of severe hypoxemia secondary to PFO associated with history of right upper lobectomy presented here, the hemithorax with volume loss and the right hemidiaphragmatic elevation might have caused RA compression with increased intracavitary pressure, and redirected blood flow from IVC to PFO. Both mechanisms would cause a right to left shunt first in the upright position and then also in recumbence, due to the displacement and compression of the RA as a result of the right hemidiaphragmatic elevation, the shunt being responsible for dyspnea and hypoxemia. The presence of polycythemia revealed a certain degree of hypoxemia that could be explained with the occurrence of POS a few months before, by the time of the pulmonary surgery.

Plateau-orthodeoxia syndrome secondary to PFO is a rare cause of dyspnea but should be considered for differential diagnosis due to its high morbidity.

It should be particularly suspected in patients with hypoxemia in the upright position, refractory to oxygen therapy, improving with recumbency, and with possible precipitating factors such as a history of lobectomy. In situations with significant anatomic distortion, right to left shunting could occur in all the positions and cause sustained severe hypoxemia, as was our case, the percutaneous closure of PFO being the only effective treatment.

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Early Detection of Left Ventricular Noncompaction in a Newborn
Left ventricular noncompaction is a cardiomyopathy characterized by excessive trabeculation of the left ventricle and deep intertrabecular recesses that communicate with the ventricular cavity. It is rarely diagnoses early in life. We report the case of a newborn, where color echo-Doppler in the first day of life showed: Left ventricular noncompaction (LVNC) due to evidence of excessive and prominent trabeculation and deep intertrabecular recesses on the left ventricular lateral wall and apex (Figure 1).

Intertrabecular recesses were filled by direct blood flow from the ventricular chamber.

Relation between LV noncompaction and LV compaction: 2.3.

Good left ventricular function.

Heart failure occurred at 14 days of life, color Doppler echocardiography showing dilatation of the left chambers, reduced systolic function, and a shortening fraction of 27%.

ECG: Right atrial overload, biventricular damage with right ventricular predominance, repolarization disturbances. Furosemide and beta-blockers were indicated, and inotropes were added at 26 days of life due to severe heart failure.

The patient died at 28 days of life due to multiple organ failure. Pathology confirmed the diagnosis: noncompaction cardiomyopathy (spongiform cardiomyopathy).

Gross examination: cardiomegaly; horseshoe kidney: small, polycystic left kidney; agenesis of the corpus callosum. Microscopical examination: myocardium with endocardial invaginations penetrating the ventricular wall thickness; endocardial fibroelastosis (Figure 2).

Left multicystic dysplastic kidney.

In 1990, Chin et al (1) described, for the first time, the isolated noncompaction of left ventricular myocardium. In 1995, the World Health Organization included it as a non-classified cardiomyopathy, and since 2006, it has been reclassified as genetic primary cardiomyopathy.

(2) According to the most widespread and accepted pathogenic theory, LVNC represents a detention in endomyocardial morphogenesis, causing excessive and prominent trabeculation and deep intertrabecular recesses.

This trabeculation often develops in the free wall and in the inferior portion of the interventricular septum, separating the ventricular inflow tract from the outflow tract. Usually between the 5th and the 8th week of gestation the myocardium matures from the