Thiamine deficiency may present four classic clinical forms: peripheral polyneuropathy, anorexia and muscular weakness (dry beriberi); high output heart failure with signs of congestion (wet beriberi); beriberi associated with shock (Shoshin beriberi) and Wernicke’s encephalopathy. In this report we describe a picture that is suggestive of severe pulmonary hypertension and cor pulmonale, with jugular stasis, congestive hepatits and generalized edema that reversed completely after the administration of thiamine.

**Case Report**

A male patient, 42, agricultural worker came to our emergency room complaining of burning epigastric pain, vomiting and watery stools. He also reported having had dyspnea at moderate effort for a long time, and which had recently worsened. He also referred having edema in both lower limbs for 15 years, which had worsened substantially in the last few days and had become generalized, including facial edema. Patient background: ex-alcoholic and smoker (one packet of cigarette – 30 years); rheumatic fever at 18, which caused double mild mitral lesion and mild aortic stenosis. Additionally, he presented a picture of high output heart failure due to beriberi in 1999, which improved after administration of thiamine. He had been using loop diuretics for 15 years to treat the edema.

On physical examination he presented blood pressure of 90x60 mmHg, respiratory rate of 24 breaths per minute, regular heart rate of 96 bpm. He presented icterus, with lower limb pitting edema and jugular stasis 3+/4+. The cardiac examination evidenced visible and palpable ictus in the fifth left intercostal space with two fingers of width, with midprecordial impulse and mid precordial impulse. The heart sounds were rhythmic, with a systolic murmur on the aortic and mitral areas 3+/6+. Pulmonary auscultation evidenced visible and palpable ictus in the fifth left intercostal area. Pulmonary hypertension. The test showed a ratio FEV1/FVC of 0.78 and FEV1 of 68%, suggestive of restriction, with no evidence of bronchodilator response. Chest tomography showed signs that suggested pulmonary hypertension and cor pulmonale, with small areas of emphysema. Investigation for HIV and schistosomiasis was negative. Pulmonary arteriography was prescribed. After three days in hospital, the patient presented cardiogenic shock, with a drop in the level of consciousness and blood pressure of 80x40 mmHg, thus requiring dopamine. Considering that the etiologic agent of the shock had not been identified, and the patient had a history of beriberi, we decided to administer 300mg of thiamine per day. After the treatment, the patient's pressure levels returned to normal, and the vasoactive medication was withdrawn after two days of treatment. With the maintenance of the diuretic medication and thiamine, the patient presented progressive improvement of the congestive picture and normalization of the hepatic enzymes (ALT=102 U/L, AST=46 U/L, bilirubins (conjugated bilirubin=0.1mg/dL and unconjugated bilirubin=0.8mg/dL) and of renal function (creatinin=0.8 mg/dL and urea=20 mg/dL).

After improvement of hemodynamic conditions, the patient underwent spirometry with a bronchodilator to investigate the pulmonary hypertension. The test showed a ratio FEV1/FVC of 0.78 and FEV1 of 68%, suggestive of restriction, with no response to bronchodilator. Chest tomography showed signs that suggested pulmonary hypertension and cor pulmonale, with small areas of emphysema. Investigation for HIV and schistosomiasis was negative. Pulmonary arteriography was negative for thromboembolism. However, surprisingly, the test evidenced pulmonary arterial mean pressure in the upper limit of normality (25 mmHg). Considering this result, a new echocardiogram was requested which was performed two weeks after the thiamine therapy had begun. The test was performed by the same practitioner that performed the first echocardiogram.
and confirmed the presence of two mild mitral lesions, but did not evidence pulmonary hypertension or dilation of the heart chambers. After twenty days in hospital, the patient was discharged with complete reversal of the congestive picture.

**Discussion**

Thiamine (vitamin B1) is a soluble vitamin that is essential for the metabolism of carbohydrates. The main sources of thiamine are cereals, grains, meat (especially pork), vegetables and dairy products. The main causes of beriberi are low intake of thiamine, alcoholism, use of loop diuretics (as they increase the renal excretion of thiamine), dialysis and use of parenteral nutrition. High consumption of carbohydrates, stress situations, such as fever and infections may aggravate the picture. The total depletion of thiamine levels in the body occurs approximately after three weeks with no supplementation.

Thiamine deficiency is classified today as an acquired error of metabolism which results in dysfunction in the Krebs cycle which would lead to ATP depletion and vasodilation with increased release of adenosine. In wet beriberi there is vasodilatation with increase of arteriovenous shunts and significant alterations in small vessels, which reduces brain and kidney circulation while increasing circulation in muscles. This phenomenon could explain alterations in the renal functions such as the ones found in our patient.

Another alteration that is characteristic of wet beriberi is the increase in end left ventricle diastolic pressure with the consequent increase in pulmonary arterial flow. These alterations could result in pulmonary hypertension. Another mechanism that could explain the pulmonary hypertension would be the increase in pulmonary vascular resistance, probably as a result of vasoconstriction. The mechanisms involved in this process have not been elucidated yet. Our patient presented a full-blown picture of cor pulmonale, with jugular stasis, congestive hepatitis and generalized edema. An aspect that is worth highlighting is the fact that cor pulmonale with manifestation of beriberi is extremely rare. For this reason, thiamine deficiency is not considered as part of the routine when investigating pulmonary hypertension pictures.

As regards the diagnosis, erythrocytic transketolase activity is the biomarker of thiamine deficiency. However, this is an expensive test that is unavailable in most medical services. This is why the therapeutic test becomes the usual way to confirm suspected cases.

Beriberi treatment consists of cardiovascular support and thiamine replacement. The precise dosage for treatment of an episode of Shoshin beriberi has not been determined yet, but daily doses of 100 to 300mg administered intravenously have been sufficient to reverse the picture. A maintenance daily dose of 100mg has been recommended. Our patient presented reversal of the cor pulmonale picture with daily doses of 300mg administered orally.

Therefore we report the case of a patient presenting a picture that was suggestive of severe pulmonary hypertension and major manifestations of right-sided heart failure including congestive hepatitis, that was completely reversed after the administration of thiamine. Some important aspects should be considered as regards our case. During the investigation there was no other factor that could explain the clinical manifestations and the course of the disease. The patient had a previous history of alcoholism and use of loop diuretic, which might explain the thiamine deficiency. Another aspect is that the patient had a previous picture of beriberi, with high output heart failure. Additionally there was immediate clinical response to thiamine replacement. Therefore, although the activity of erythrocytic transketolase had not been dosed, beriberi arose as the most likely hypothesis for our patient. Finally, our report presents a limitation that is worthy of notice. The presence of high output syndromes puts a limit on the diagnosis of pulmonary hypertension using the echocardiogram, since the echocardiographic variables can change due to high output heart failure itself. This is why the definitive diagnosis of pulmonary hypertension cannot be established without confirmation by an invasive hemodynamic study.

To conclude, we believe that this case is very important since thiamine deficiency can be considered as a reversible cause of pulmonary hypertension. Therefore, beriberi should be included for differential diagnosis purposes in patients with cor pulmonale/right-sided heart failure.

**Potential Conflict of Interest**

No potential conflict of interest relevant to this article was reported.

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**Study Association**

This study is not associated with any graduation program.

**References**