BERIBERI HEART DISEASE IN TWO THAI LABORERS WORKING IN SAUDI ARABIA: RECOVERY FROM SHOSHIN BERIBERI
MAHESH C. GARG, MRCP; V ASUDEVAN SIVANANDAM, MD; MOHAMMED Z. KHAWAJI, MD, PAULO A. RIBEIRO, MD

MORE THAN 200 cases of unexplained sudden death, in apparently healthy Thai workers, have been reported in the Far East during the last year. Described herein are two patients with beriberi heart disease. Both patients presented with a recent history of dyspnea, ankle edema, tachycardia, and with physical signs of biventricular heart and clinical features of high cardiac output failure. The diet regimen was low in thiamine and both patients ate polished rice three times a day. Echocardiographic and Doppler studies revealed, in both cases, a left ventricle with a normal size and ejection fraction. The right atrium and ventricle were dilated. Despite conventional anti-failure therapy with digoxin, diuretics, and eventually intravenous inotropic support, one of the patients developed severe cardiogenic shock and metabolic acidosis. There was a dramatic hemodynamic response to intravenous thiamine and the patient recovered within hours. In both cases, the clinical features of high cardiac output status with biventricular heart failure subsided with thiamine therapy. These findings were corroborated by the normalization of the heart size and cardiac output in both cases.

Case Reports

Patient 1
A 33-year-old Thai male laborer who had been working for two years in Saudi Arabia, presented with a 9-day history of dyspnea, orthopnea, abdominal pains, and marked ankle edema. His past medical history was unremarkable. He had been on a diet of three daily meals consisting of polished rice, eggs, and chicken. On examination the jugular venous pressure was 15 cm, blood pressure was 130/60 mm Hg, and the heart rate was 94 beats/min and regular. Ascultation of the chest revealed bilateral basal crepitations. There was an ejection murmur (grade 2/6) at the left sternal border. The patient had a striking gallop rhythm. The femoral artery pulses were collapsing. The liver was 3 cm below the costal and was not pulsatile. There was bilateral pitting ankle edema with warm extremities. The renal profile was normal but there was a slightly raised bilirubin and SGOT. Pyruvate level on arrival was 320 mmol/L (normal, 20 to 50). Thyroid function tests were normal. The electrocardiogram revealed sinus rhythm with nonspecific ST-T changes. The chest x-ray showed cardiomegaly with a cardiothoracic ratio of 60%. There was right pleural effusion and pulmonary congestion. A cross-sectional echocardiogram was done and showed that the left ventricular size was 5.2 cm in diastole and 3.4 cm in systole, and there was trivial mitral and tricuspid incompetence. The right atrium and right ventricle were markedly dilated. Doppler cardiac output measured 9.6 L/min. Conventional anti-failure therapy was instituted, and 48 hours after admission the patient became confused and tachypneic. The patient was clinically in left ventricular failure and was started on inotropic support with dopamine and dobutamine. Over the next few hours the patient deteriorated further and went into cardiogenic shock and developed severe metabolic acidosis; the blood pressure was 58/40 mm Hg with a pH of 6.9. At that point, intravenous thiamine 100 mg was started every 8 hours.

From the Department of Medicine (Drs. Garg, Sivanandam, and Khawaji), Riyadh Central Hospital and Department of Cardiovascular Diseases (Dr. Ribeiro), King Faisal Specialist Hospital and Research Centre, Riyadh, Saudi Arabia.

Address reprint requests and correspondence to Dr. Ribeiro: Department of Cardiovascular Diseases, MBC 16, King Faisal Specialist Hospital and Research Centre, P.O. Box 3354, Riyadh 11211, Saudi Arabia.

Journal of the Saudi Heart Association, Vol. 4, No.1, 1992
and the metabolic acidosis was corrected. His recovery was dramatic and within two hours he was talking and sitting on the bed. Over the next few weeks thiamine therapy was continued. The cardiac output measured two days after discharge from the coronary care unit was 5.7 L/min and at hospital discharge was 4.6 L/min. The decrease in cardiac output correlated with the patient’s marked clinical improvement. The chest x-ray demonstrated that the cardiac size had normalized.

**Patient 2**

A 40-year-old Thai male was admitted to the hospital with a 5-month history of paraesthesia in the hands and feet and with a 2-week history of atypical chest pain and shortness of breath on exertion. Two days prior to admission, the patient experienced further deterioration and was orthopneic. His past medical history was unremarkable with no history of diabetes, hypertension, or alcoholic intake. His diet was based on three meals of polished rice. On physical examination his jugular venous pressure was up 15 cm; sinus rhythm, 85 beats/min; and blood pressure, 140/70 mm Hg. The chest was clear and there was an ejection systolic murmur (grade 2/6) at the sternal border. The liver was not palpable and his femoral pulses were collapsing in character. There was mild ankle edema and hot extremities. His electrocardiogram showed sinus rhythm, normal axis, and nonspecific ST-T changes. His blood tests included a full blood count, and renal profiles were normal with an SGOT of 67, which remained persistently high throughout his admission. A chest x-ray showed bilateral pleural effusions and a cardiothoracic ratio of 55% with a pulmonary edema (Figures 2A, 2B). Cross-sectional echocardiogram revealed a normal-sized left ventricle with a diastolic dimension of 5.5 cm and systolic of 3.2 cm and an ejection fraction of 73%. The cardiac output measured 8.4 L/min on Doppler. There was mild-to-moderate tricuspid incompetence and a markedly dilated right atrium and right ventricle. There was no atrial septal defect on Doppler. He was started on parenteral thiamine, 100 mg intravenously every 8 hours, and 24 hours later he was switched to oral thiamine. Conventional anti-failure therapy was instituted with digoxin and diuretics. The physical signs of high cardiac output failure subsided within 1 week and the cardiac output decreased to 4.5 L/min. The chest x-ray showed a marked reduction of the cardiac size and normalization of the lung fields (Figures 2A, 2B). A repeat echocardiographic study showed normalization of the right atrium and right ventricular size. Prophylactic dietary instructions were given to both patients before hospital discharge.
BERIBERI HEART DISEASE

Figure 2. Chest x-ray before (A) and after treatment with thiamine (B) in patient 2. Note the decrease in heart size and clear lung fields after therapy with thiamine.

Discussion

The reappearance of beriberi heart disease has been reported in 23 young Japanese patients. These were teenagers on a diet with thiamine deficiency. Conceivably, many Thai patients who have recently experienced sudden death in the Middle and Far East developed Shoshin beriberi. Both of our Thai patients were on a low thiamine diet, and one of them would have died if not given intravenous thiamine. Shoshin beriberi leads to a fulminant death after the development of metabolic acidosis, high pyruvate levels, pulmonary edema, and ultimately cardiogenic shock.

The clinical and hemodynamic response to thiamine after the administration of intravenous thiamine was dramatic. This type of beriberi was responsible for thousands of deaths in Japan before the discovery of thiamine deficiency. Both of our Thai patient were on a diet based on polished rice and were deficient in thiamine. An investigation is currently being undertaken in the labor camps where these patients lived.

In the Western world, Shoshin beriberi is very rare. Beriberi heart disease is usually associated with chronic alcoholism, particularly in beer drinkers because the high carbohydrate content of beer increases the demand for thiamine. These findings should stimulate investigating cases of sudden death occurring in Thai laborers in the Middle East for possible thiamine deficiency.

The cardinal clinical features of these patients were those of a high cardiac output status with hot extremities. These physical signs disappeared after administration of thiamine. Patients with Shoshin beriberi develop severe metabolic acidosis and this may explain the high level of pyruvate found in this condition. Peripheral vasodilation and low systemic resistance with high ventricular filling pressures contribute to the high cardiac output state. Both our patients presented with predominantly right heart failure. Both the right atrium and right ventricle were dilated and the left ventricular size and function were normal.

We measured the pyruvate blood levels on the day of arrival and indeed they were extremely high in patient 1. The laboratory diagnosis is made by the demonstration of increased levels of pyruvate and lactate and the presence of a low transketolase level. The best test is the increase in erythrocyte transketolase with therapy. We demonstrated a very high level of pyruvate, but the transketolase sample sent abroad was lost. In both cases there was a high cardiac output state that was corrected with the administration of parenteral thiamine. We also excluded other causes of high cardiac output state such as thyrotoxicosis, Paget's disease, arteriovenous fistulas, and alcoholism.

The cornerstone therapy in beriberi is thiamine.
However, the acute reversal of the vasodilatation induced by correction of this deficiency may precipitate the unprepared ventricle to go into a low output state. Patients should therefore receive anti-failure therapy with digoxin and diuretics, concomitantly with administration of thiamine. The correction of metabolic acidosis has to be done in patients with Shoshin beriberi and is of paramount importance for the patient's survival.

This paper describes two cases of beriberi heart disease of Thai laborers in the Middle East. One of our patients had the Shoshin form of beriberi, and intravenous treatment with thiamine reversed the patient's cardiogenic shock. This finding may indicate that many of the sudden deaths occurring in Thai laborers in the Middle and Far East maybe due to beriberi heart disease.

References