Case Report

Pediatric Cardiac Beriberi: 3 Different Presentations

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The authors report 3 cases of pediatric cardiac beriberi at Queen Sirikit National Institute of Child Health during the last 10-year-period. The first two cases presented classically while the third case came with an unusual presentation. Cardiac beriberi is an old disease in modern Bangkok which can present in as several different clinical patterns. The diagnosis needs a high index of suspicion. Echocardiographic findings of right heart dysfunction and dramatic response to intravenous thiamine are diagnostic.

Keywords: *Cardiac beriberi, Failure, Shock, Cyanosis, Thiamine, Bangkok, Thailand*

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Cardiac beriberi has been described to be associated with thiamine deficiency for centuries[1]. It is common where highly polished rice is the staple diet[1-3] and other primary dietary sources of thiamine (meat, fish, and legumes) are in short supply[4]. Thiamine deficiency is compounded when local diets include foods such as fermented fish[5], tea leaves, and betel nut, which contain antithiamine factors[6,7]. Once, it was a major cause of infant mortality in the north-western border of Thailand[8]. Now, in the modern era, the disease is rarely diagnosed. Queen Sirikit National Institute of Child Health (QSNICH) is a tertiary care hospital with about 1000-1500 new cardiac cases/year. The diagnosis of cardiac beriberi at QSNICH during the last decade has been entertained in 3 cases with different presentations which will be reviewed in the present study.

Case Report

Case 1

A one-month-old baby had weak cry and edema for one day. Two days before admission, she had pass loose stool 3-4 times/day together with occasional vomiting. Additional history revealed that she was the first born child in the family, exclusively breast-fed. Her mother's staple diet included polished rice with only small amounts of meat, fish, and legumes intake. She had no history of weakness or paresthesia. At the emergency room, the baby was immediately intubated and resuscitated for cardiopulmonary failure. Physical examination revealed a 4.4 kg-baby, afebrile, drowsy, mild generalized edema, hypotonia, heart rate 168/minute, unmeasurable blood pressure, increased pulmonic component of second heart sound, no heart murmur, hepatomegaly and hyporeflexia. Chest X-ray revealed mild cardiomegaly with pulmonary congestion. Electrocardiogram revealed prolonged P-R and Q-T intervals. Echocardiogram revealed no structural defects, right atrial enlargement, right ventricular enlargement with poor right ventricular function, mild tricuspid regurgitation with estimated systolic pulmonary pressure of 45 mmHg. The left ventricular function was normal. After 50 mg intravenous thiamine administration, the baby's condition improved rapidly and she was successfully extubated with complete resolution of all abnormalities after 6 hours.

Case 2

A 12-year-old boy was referred for treatment of suspected myocarditis. He was previously well until

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1 week before admission when he felt calf pain, became edematous and developed paresthesia of legs followed by progressive dyspnea. At the referring hospital, he was diagnosed with myocarditis and treated with dobutamine together with furosemide. Additional history revealed that the boy was Islamic and during the religious fasting period, had taken only polished rice for months. Physical examination revealed a 50-kg-boy, good consciousness, mildly edematous legs, heart rate 146/minute, blood pressure 106/65 mmHg, body temperature 37.1°C, respiratory rate 28/minute, normal heart sounds and no heart murmur. Neurological examination revealed generalized hypotonia with hyporeflexia. Chest X-ray revealed mild cardiomegaly with pulmonary congestion. Electrocardiogram revealed sinus tachycardia with generalized low voltage waves. Echocardiogram revealed no structural defects, right atrial and right ventricular enlargement, moderate tricuspid regurgitation with estimated systolic pulmonary pressure of 43 mmHg and normal left ventricular function. After 30 mg intravenous thiamine administration for 2 hours, he had diuresis and rapidly improved. On the following day, all abnormalities completely resolved.

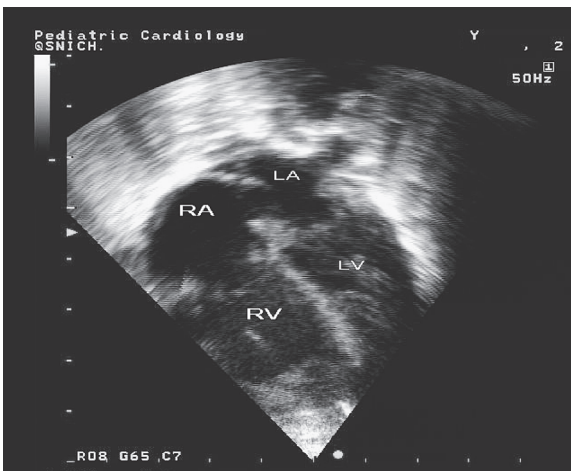

Fig. 1 Echocardiogram revealed right atrial and right ventricular enlargement (RA = right atrium, RV = right ventricle, LA = left atrium, LV = left ventricle)

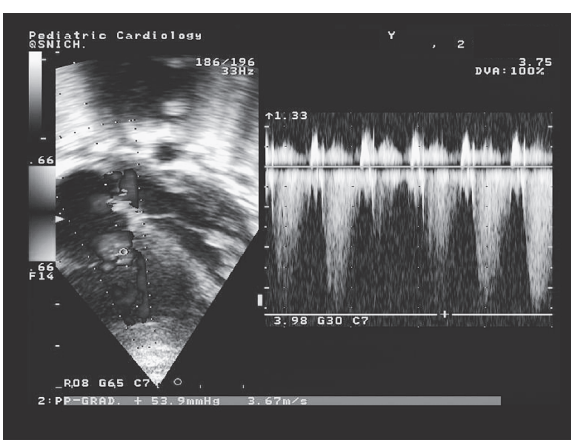

Fig. 2 Echocardiogram revealed moderate tricuspid regurgitation with estimated systolic pulmonary pressure of 60 mmHg

Case 3

A one-month-old baby presented with a weak cry for one day. She was the seventh offspring of the family, exclusively breast-fed; previously well with an uneventful prenatal and delivery history. Additional history revealed that her mother had been experiencing paresthesia of hands and feet for years, taking polished rice as her major diet with minimal meat, fish, and legumes. At the emergency room, she was given assisted ventilation with 100% oxygen. Physical examination revealed a 5.3 kg-baby, drowsy, with central cyanosis, hypotonia, heart rate 150/minute, blood pressure 74/50 mmHg, body temperature 37.0∞C, no heart murmur, increased pulmonic component of second heart sound, hepatomegaly and hyporeflexia. Her arterial partial pressure of oxygen was 34 mmHg. Chest X-ray revealed mild cardiomegaly with pulmonary congestion. Electrocardiogram revealed prolonged Q-T interval. Echocardiogram revealed no structural defects, right atrial and right ventricular enlargement, right-to-left shunting via the foramen ovale, mild mitral regurgitation, moderate tricuspid regurgitation with estimated systolic pulmonary pressure of 60 mmHg, poor right ventricular function with normal left ventricular function (Fig. 1, 2). After 50 mg intravenous thiamine administration for 6 hours, the patient was successfully extubated with complete resolution of all abnormalities (Fig. 3).

Discussion

The diagnosis of cardiac beriberi has been demonstrated in three cases at QSNICH during this last decade (August 1999, October 2005 and December 2007). All 3 cases presented with signs of congestive heart failure or shock in the absence of fever or other signs of sepsis, hypovolemia or structural cardiac abnormalities. All also responded rapidly with dramatic clinical improvement following parenteral thiamine

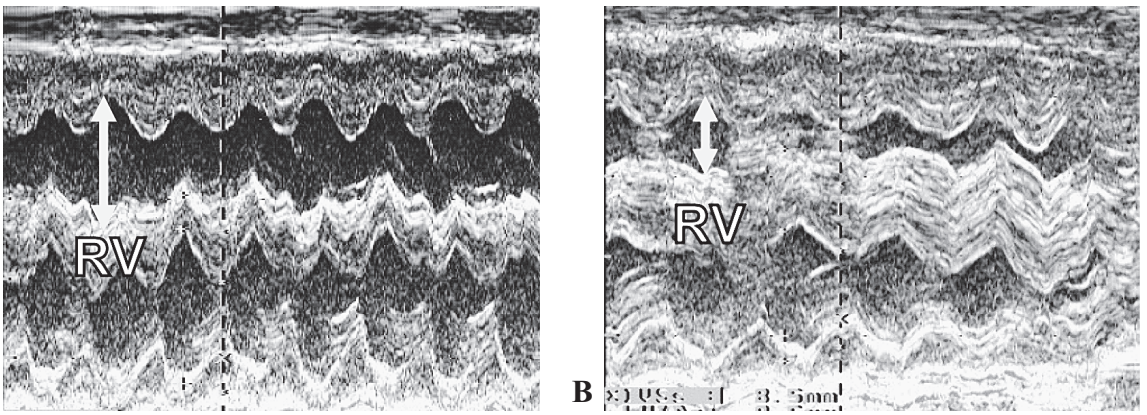

Fig. 3 M-Mode echocardiogram revealed right ventricular (RV) dilatation on 3A which resolved 6 hours later (3B)

administration[9]. The first two cases are classic cases of cardiac beriberi (infantile type and adult type respectively)[10]. Cyanosis in the third case is an unusual presentation which has never been reported before.

A history of neurological symptoms in children or in mothers is helpful for diagnosis. In the present study, this was encountered in case 2 (child) and case 3 (mother). Precise neurological examination is helpful to raise higher indices of suspicion in all, especially for hyporeflexia. However, the correlation between thiamine deficiency and the presence of clinical signs thought to indicate thiamine neuropathy is poor[1].

Clinically, significant thiamine deficiency in breast-fed infants has been shown to relate to methods of cooking rice, the food selected by lactating mothers and the family's socioeconomic status[9]. All of the cases in the present study go along with previous published studies. Biochemical studies are helpful for the diagnosis but not necessary[3,9].

Color Doppler echocardiogram is helpful for diagnosis. An echocardiographic study in a patient with cardiac beriberi was first reported in 1990 showing isolated right heart dilatation and tricuspid regurgitation detected by Doppler[11]. The estimated pulmonary pressure in the present report is similar to previous studies[11,12]. Right-to-left shunt via the foramen ovale demonstrated by Color Doppler echocardiogram in the third case explains the cyanosis mimicking cyanotic congenital heart disease. The most important clue for the diagnosis is the dramatic response after intravenous thiamine administration with complete resolution of cardiac dysfunction.

Dr. Edward Bright Vedder (1878 - 1952), a U.S. Army physician, began experimenting with the treatment of infantile beriberi with an extract of rice polishings (or partially milled rice, an alcohol-based extract of rice hulls)[13]. Vedder and Chamberlain cured 15 infants whose mothers had symptoms of beriberi by supplementing each mother's milk with an extract of rice polishings and allowing nursing to continue[13]. The mortality in infants with acute infantile beriberi has decreased significantly from 250 to 78 deaths per 1000 live births with the introduction of intramuscular thiamine (50 mg)[14].

The dosage varies from study to study, with as high as 500 mg intravenous thiamine administered in adult patients[15]. In the present report, the second case improved rapidly with only 30 mg thiamine intravenously.

Conclusion

Cardiac beriberi is an old disease in modern Bangkok which can present with several clinical patterns. The diagnosis needs a high index of suspicion. Echocardiographic findings of isolated right heart dysfunction and dramatic response to intravenous thiamine are diagnostic.

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โรคหัวใจที่เกิดจากการขาดวิตามินบี 1: รายงานผู้ป่วย 3 ราย

ชัยสิทธิ์ แสงทวีสิน, สมศักดิ์ เลิศวีรวัฒน์

คณะผู้รายงานนำเสนอรายงานการรักษาผู้ป่วยโรคหัวใจที่เกิดจากการขาดวิตามินบี 1 ที่สถาบันสุขภาพเด็กแห่งชาติมหาราชินี จำนวน 3 รายในรอบ 10 ปีที่ผ่านมา รายแรก และรายที่สองเป็นตัวอย่างผู้ป่วยที่ตรงตามตำราโดยทั่วไป ส่วนรายสุดท้ายมาด้วยอาการที่ไม่เคยมีรายงานที่ใดมาก่อน โรคนี้ยังพบได้อยู่แม้แต่ในกรุงเทพฯ ที่ทันสมัยขึ้นมากแล้ว การจะวินิจฉัยได้จำเป็นต้องนึกถึงโรคนี้ไว้ด้วยเสมอ การตรวจคลื่นเสียงสะท้อนหัวใจที่พบการทำงานที่ผิดปกติเฉพาะหัวใจด้านขวา และการตอบสนองที่ดีต่อการให้วิตามินบี 1 ทางหลอดเลือดดำช่วยสนับสนุนการวินิจฉัยเป็นอย่างดี