Diagnosing Beriberi in Emergency Situations

by Prof Mike Golden, Aberdeen University.

This piece will be most useful to medical professionals - doctors and nurses. However, the article may also be useful for non-specialists who need to communicate with medical personal about this deficiency disease. In some places medical terms have been simply explained for the non-specialist.

Recently the NGO community in West Africa learned of an outbreak of beriberi in an area with severe nutritional deprivation. The diagnosis had been based upon the presence of epidemic oedema found during a nutritional survey. The survey team included experienced, doctors. The prevalence of beriberi was estimated at 30%. I accompanied an ACF team working in the area to examine these patients. The only clinical feature that these patients had that resembled beriberi was the oedema. The patients had 'famine oedema' which is the same as adult kwashiorkor. In the absence of diagnostic laboratories, the diagnosis of nutritional deficiency diseases depends upon:

1. a high index of suspicion;
2. knowledge of the likely deficiencies in different circumstances; and
3. the clinical skills, knowledge and experience to elucidate the characteristic clinical features of a deficiency.

Clinical ability is the most important. This paper addresses beriberi which is primarily due to thiamine (vitamin B₁) deficiency. Most forms of beriberi are easily confused with other illnesses, especially as a mixture of syndromes may be present. The condition is frequently missed in clinical practice unless the text book symptoms of wet, dry or aphonic beriberi are present. My recent experience in West Africa confirms the need to strengthen diagnostic capacity for this condition. Clearly there is a need for additional training material which can be used by people in the field. A first step in developing this material may be to encourage health and nutritional workers to take pictures and videos of affected populations when outbreaks of beriberi are confirmed. In addition it is important to include descriptions of circumstances under which a particular beriberi problem is arising specifying demographic and nutritional characteristics of affected populations.

This short article attempts to set out when to suspect beriberi, the different syndromes associated with the deficiency disease and how to treat the condition.

When to suspect beriberi.

Beriberi occurs where there is a high carbohydrate or alcohol intake and a low thiamine intake. This classically occurs amongst populations consuming polished non parboiled rice, particularly where the rice is contaminated with moulds. It also occurs in alcoholism. However, raw fish and some ‘bush’ teas or vegetables contain anti-thiamine enzymes that can precipitate beriberi. The condition therefore usually occurs where energy intake is good and energy expenditure high - beriberi is not a feature of starvation.

Types of beriberi.

Beriberi causes cardiovascular and neurological signs and symptoms. There are eight clinically recognisable syndromes - five in adults and three in children. Often a patient has features of more than one of the syndromes. Beriberi bears no relationship to anthropometric status - fat people get beriberi and it also occurs in the fully-breastfed infant.

Wet beriberi
The resistance between the arteries and the veins drops to such an extent that the blood flows round the body much more rapidly than normal, the heart is unable to maintain this high output and fails.

These patients have oedema which needs to be distinguished from:

- famine oedema (adult kwashiorkor);
- nephritis (kidney inflammation);
- nephrotic syndrome (kidney disease); and
- intoxication (heavy metals, ethylene glycol, etc.).

Patients may also have hyperdynamic circulation and a large heart with the following signs:

1. Restlessness and arms and legs feel warm to touch.
2. Pulse pressure is high with an increase in the systolic pressure and a decreased diastolic pressure (e.g. 140/40); when the wrist is encircled lightly the pulse is easily felt (water hammer pulse); slight pressure on the nails so that they go white shows pulsation in the nail bed.
3. The jugular venous pulse (vein pulse in the neck) is raised with pronounced pulse waves.
4. The apex of the heart is displaced to the left so that it may reach the axilla and it is easily felt; there are usually systolic 'flow' murmurs and a third heart sound giving a 'galloping' rhythm that is accentuated when the patient lies on the left side and breathes in.

Unlike other forms of heart failure the patient's lungs are usually clear. They can lie flat on their backs without getting breathless and they are not cyanosed.

The hyperdynamic circulation could also be caused by:

1. severe anaemia;
2. thyrotoxicosis (hyperthyroid);
3. pregnancy;
4. chronic liver disease;
5. strongyloidiasis (worm infestation); and
6. arteriovenous fistulae (a rare arterial malformation).

**Shoshin beriberi**

The heart is primarily affected and these patients have classical heart failure and sudden death. These patients are more breathless when they lie down and have crepitations (wet crackles) in their lower lung fields. The incidence of Shoshin beriberi in areas with an outbreak of wet beriberi is unknown because it is not recognised and the death ascribed to some other cause of heart failure.

**Dry beriberi**

(this is the form that has been experimentally induced in human volunteers). In dry beriberi there is a peripheral neuritis. It starts with paraesthesia (pins and needles) of the feet, diminished touch sensation, and a feeling of 'heat' in the feet. Joint position, vibration and pain sensation are usually normal. The ankle and then the knee reflexes are lost and the patients have muscle weakness starting with the foot.

**Korsakoff's psychosis**

(common in alcoholics) This is characterised by profound loss of recent memory, with preservation of past memory and an active imagination. These patients may concoct very plausible stories about what they themselves (not other people) have been doing, to cover their memory defect and do not admit to any loss of memory (called confabulation). It is necessary to know what the patient has actually been doing before the history is taken. The defect is irreversible with thiamine treatment, but its progression is stopped.

**Wemickie's encephalopathy**

(Alcoholic encephalopathy)
These patients start with irritability and forgetfulness and progress to having characteristics of damage or destruction of the small organs on the base of the brain (mamillary bodies) that control eye movements. Symptoms include:

- drooping of their eye lids (ptosis), and squints where the eye does not move outwards (ophthalmoplegia),
- horizontal flickering of the eyes (nystagmus). Other cranial nerve lesions,
- the patients may have an unsteadiness of their hand and foot movements (cerebellar ataxia);
- progressing to confusion and delirium before death.

Minor forms of this type of beriberi are quite common - up to 10% of post-mortem examinations in Australia show damage to the mamillary bodies. These patients are often misdiagnosed as having other forms of encephalopathy (brain inflammation) such as viral illness, cerebral malaria, sleeping sickness, etc.

**Infantile acute cardiac beriberi**

The peak prevalence of this form occurs in fat breastfed babies of 1-3 months of age. The first signs appear like colic with screaming bouts, restlessness, anorexia and vomiting. This progresses to oedema, cyanosis and breathlessness with signs of heart failure:

- increased pulse rate;
- enlargement of the heart;
- additional heart sounds;
- a systolic murmur;
- pulmonary oedema;
- liver enlargement and low urine volume.

The children die from heart failure: the correct diagnosis is rarely made. Typhoid, malaria, pneumonia and septicaemia are commonly confused with this form of beriberi.

**Aphonic beriberi**

The peak prevalence is in 4-6 month old children. The child's voice changes so that the cry gets more and more hoarse until no sound at all is produced. Strikingly, these children do appear to be crying without any noise. Without treatment they progress over a few days to restlessness, oedema, breathlessness and then death.

**Infantile encephalitic beriberi**

(sometimes called 'pseudомeningеal' beriberi)

This usually occurs in 6-12 month old children and is the equivalent of Wemickie's encephalopathy in the infant. The patient appears to have encephalitis! meningitis. Signs include:

- nystagmus (flickering of the eye),
muscle twitching; a bulging fontanelle; and convulsions and unconsciousness.

When a lumbar-puncture is done the cerebro-spinal fluid appears to be 'gin' clear and does not froth easily when it is shaken. This can be confused with all forms of encephalitis and meningitis, malaria and even acute vitamin A intoxication. Older children get the same features as adults.

**What to do when beriberi is suspected.**

After you have carefully examined the patient and described the findings in all the systems, then you should give 'flooding doses' of thiamine (normal adult requirement is 0.4mg per 1000 kcal). Give 50 or 100 mg thiamine hydrochloride intravenously and then give 10 mg per day orally. Infants may be treated with adult doses (it is not toxic in very large quantities), or lesser doses if this is more convenient. Mothers of patients should always be given 10mg per day orally.

The response is rapid in wet and infantile beriberi. A diuresis quite suddenly starts between 4 and 48 hours after the thiamine is given (it is unclear why the delay occurs). It is dramatic with visible resolution of much of the oedema over 4 to 8 hours followed by complete loss over the next 48 hours. Shoshin and infantile cardiac beriberi also respond dramatically with resolution of the heart failure. The convulsions and delirium in encephalitic patients respond rapidly and the patient's life is saved. However, the cranial nerve lesions can take months to recover and may be permanent. Aphonia gradually resolves over two to three weeks. The mental changes is Korsakoff's psychosis are permanent and do not respond to treatment.

In population surveys urinary thiamine levels of less than 65 ug/g creatine indicates thiamine deficiency but this cannot be used in individual diagnosis as urinary thiamine levels in individuals vary so markedly over time (high coefficients of variation). Text books can be consulted for other biochemical tests which are appropriate for individual diagnosis.

We asked Annalies Borrel, nutritionist with Concern Worldwide, to comment on this article.

She said that "the subject is most relevant and addresses a very real query in the field". She recalled being in Angola and coming across just this problem. Beriberi was suspected in a child but no-one was sure how to distinguish it from Kwashiorkor, how to confirm the diagnosis and how to determine whether there was a population level problem and if so how severe it was. Annalies felt that the article highlighted the need to develop user-friendly guidelines to help field staff distinguish between oedema due to beriberi and oedema due to Kwashiorkor. She also felt it important to make the point that the existence of individual cases of beriberi often reflects food related problems at population level and that a focus on clinical diagnosis and treatment leaves out an important set of issues - namely, how do we assess, address and respond to a population level problem of beriberi?

Taken from Field Exchange 1 www.ennonline.net/fex/1/diagnosing

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