Beriberi

Synonyms: thiamine deficiency, aneurin deficiency, vitamin B1 deficiency

Beriberi is caused by deficiency of thiamine, also called aneurin and vitamin B1. This is a water-soluble, heat-labile vitamin that acts as a coenzyme on a number of metabolic pathways. The recommended daily intake is 1.2 mg.\(^\text{[1]}\) The cardiovascular, muscular, gastrointestinal and nervous systems can be affected.

The word 'beri' means weak in Sinhala, a language from Sri Lanka. Beriberi means very weak. Another translation is 'I cannot, I cannot'.\(^\text{[2]}\)

Epidemiology

Beriberi was endemic in some areas of the world and may be related to the consumption of milled rice. The process removes the outer crust that contains the vitamin and, in Indonesia especially, the condition was very common. The disease tends to affect adults and infants between 1 and 4 months old. Improved nutrition, health propaganda and heightened public awareness all contributed to a reduction in prevalence since the 1980s.\(^\text{[3]}\)

Outbreaks occasionally occur in circumstances in which there are dietary restrictions and/or intake of thiamine antagonists.\(^\text{[4, 5]}\) An outbreak in Israel in 2003 was traced to the use of soy-based milk formula.\(^\text{[6]}\)

Risk factors

In western societies, predisposing factors to thiamine deficiency include gastrointestinal disease and alcoholism, as the latter impairs absorption of the vitamin. Deficiency has been recorded in AIDS, after bariatric surgery and in hyperemesis gravidarum.\(^\text{[7, 8]}\) It can occur (rarely) after any type of protracted vomiting state. Cases have been reported in which severe vomiting was associated with post-cholecystectomy and suspected Crohn's disease.\(^\text{[9, 10]}\) Hypermetabolic states also predispose to deficiency and this includes fever, pregnancy, postoperative state, total parenteral nutrition and renal dialysis.\(^\text{[11]}\) A loop diuretic can also increase the risk.\(^\text{[12]}\)

Presentation

History

Early features

- Fatigue and apathy.
- Irritability.
- Drowsiness, depression, poor concentration.
- Anorexia, nausea, vomiting, abdominal pain.

Later features

- Paraesthesia, peripheral neuropathy, depressed tendon reflexes, loss of vibration sense.
- Tender leg muscles and muscle cramps.
- Congestive heart failure with dyspnoea, orthopnoea and oedema.

In infancy, beriberi can present as acute fulminating, acute cardiac, aphonc and pseudomeningitic. In the aphonc form there is hoarseness and a weak cry due to paralysis of the laryngeal nerve. In the pseudomeningitic variety there are symptoms of nystagmus, vomiting and convulsions but normal cerebrospinal fluid (CSF).
Examination
Beriberi is often classified as 'dry beriberi' when features are neurological and muscular and 'wet beriberi' when there is predominantly heart failure. Physical findings include:

- Pallor and waxy skin.
- Signs of malnutrition and wasting.
- Listlessness, weakness.
- Tachycardia, hepatomegaly, cardiomegaly and peripheral oedema.
- Paraesthesia and peripheral neuropathy, including depressed tendon reflexes.

Two conditions caused by thiamine deficiency that are usually related to alcohol abuse in western societies are Wernicke's encephalopathy and Korsakoff's syndrome. See separate article Wernicke-Korsakoff Syndrome.

Diagnosis
A high index of suspicion should be maintained for patients with chronic alcohol abuse, special diets and malnutrition.

Congestive heart failure may need to be excluded as may other vitamin deficiencies (vitamin B12, niacin) if neurological symptoms are present.

Investigations

- Probably the best diagnostic test is a good clinical response to administration of intravenous thiamine.
- Another test (performed occasionally as it is expensive) is a thiamine loading test. This involves administering a test dose of thiamine. In deficient patients the urine excretion of thiamine and thiamine pyrophosphate is lower than normal. Pre-loading and post-loading whole blood or erythrocyte transketolase activity is also sometimes assessed.
- Other tests include blood thiamine level, pyruvate, lactate, alphaketoglutarate and glycosylate.
- Thiamine concentration in whole blood can now be measured using high-performance liquid chromatography.
- In Wernicke's encephalopathy, the CSF is normal or protein is slightly elevated but, if left untreated, blood pyruvate and blood transketolase will rise.
- MRI scan and, to a lesser extent CT, have been used to detect brain changes in Wernicke's encephalopathy.
- The electroencephalogram (EEG) may show diffuse slowing.
- Echocardiogram should be used to assess heart failure.

Management
There is often a deficiency of more than one vitamin and so the patient must be carefully monitored as treatment starts.

Thiamine is available in both oral and parenteral form and the latter may be employed at first, with transfer to oral preparations later. However, the Medicines and Healthcare products Regulatory Agency (MHRA) advises that potentially serious allergic adverse reactions may occur during, or shortly after, parenteral administration. They recommend that:

- Use be restricted to patients in whom parenteral treatment is essential.
- Intravenous injections should be administered slowly over 10 minutes.
- Facilities for treating anaphylaxis should be available.

25-100 mg daily by mouth are recommended for chronic mild deficiency and 200-300 mg daily for more severe deficiency.

Vitamin B deficiency, other than vitamin B12 deficiency, is rare in the UK and should be treated with preparations containing thiamine (B1), riboflavin (B2) and nicotinamide. Parenteral administration followed by oral treatment are recommended for Wernicke's encephalopathy and Korsakoff's syndrome.

There is usually no need for the usual diuretics and angiotensin-converting enzyme (ACE) inhibitors for heart failure as there is often improvement in 6 to 24 hours and marked improvement in a few days. However, some patients can go into standard cardiac failure and require input from a cardiologist.

If alcoholism is the underlying problem, a 3-stage approach is required:

- If alcohol is suddenly stopped, a withdrawal syndrome of delirium tremens may result. A reducing dose of benzodiazepine, usually chlordiazepoxide, should therefore be started during the withdrawal process, the starting dose being dependent upon the level of alcohol consumption.
- The next stage is to get the patient to admit that there is a problem of alcohol abuse.
- The final stage is to get the patient to enter into long-term management of the problem. For more information, see separate article Alcoholism and Alcohol Abuse - Management.

Complications
- The heart failure of beriberi, if untreated, can be fatal.
- Anaphalaxis can result from parenteral treatment.
Prognosis

Mortality is rare and is usually associated with the wet form due to cardiac failure. Morbidity is also rare and usually presents in the dry form with neurological symptoms. In both situations, recognition of the condition and early replacement with thiamine rapidly reverse organ dysfunction. Korsakoff's syndrome may, however, only be reversible to a small degree.

Prevention

Adults or the families of children need education about the cause of the disease and how to prevent recurrence. In third world countries the child may be discharged back to the same environment.

Good dietary sources of thiamine are whole grains, lean pork and legumes. Thiamine is not present in fats, oils and refined sugars and is destroyed by heat, pasteurisation and ionising radiation. Freezing does not affect thiamin content of foods.

In developed countries the problem is usually one of the very difficult management of alcohol abuse. If there are other reasons, such as after bariatric surgery, supplements can be continued.

History

The disease was described by the Chinese around 2700 BC but the cause was unknown. In the 19th century it was realised that the diet needed protein, fat and most of the calories from carbohydrate but it was not until the late 19th and early 20th centuries that the need for other nutrients was appreciated. Christiaan Eijkman studied prisoners in the Dutch East Indies who were fed a diet of polished rice and he noted a high incidence of neurological disorders. He also found that either feeding prisoners the whole rice or adding the husk would reverse the disorder. In 1911 Funk confirmed such findings in birds and postulated substances called vital amines, later abbreviated to vitamins.

Beriberi was not uncommon on long naval voyages and was thought to have much in common with scurvy. In 1753 Royal Navy Surgeon James Lind demonstrated that the consumption of fresh lemons and oranges cured scurvy.

In 1926 two biochemists, called Jansen and Donath, isolated a tiny amount of a substance they called aneurin but there was too little to be of much value.

In 1928, a young Japanese medical officer, Kanehiro Takaki, arranged that twelve sailors into four groups of three to try three treatments and a control group in what was almost the first randomised controlled trial on record. There was a similar naval controlled trial in the early 1880s. A young Japanese medical officer, Kanehiro Takaki, arranged that two ships left Japan on similar voyages but with different diets. The first ship served the usual fare of rice, with some vegetables and fish. The second also served the crew wheat and milk, in addition to more meat than was served on the first ship. The results were impressive with 25 deaths from beriberi on the first ship and none on the other. The Japanese Admiralty adopted the new diet for the entire navy.

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