

This factsheet describes Wernicke-Korsakoff syndrome and related conditions. It also looks at the service and treatment needs of people affected by this syndrome

Wernicke-Korsakoff syndrome

Wernicke-Korsakoff syndrome (WKS) is a form of brain damage associated with alcohol misuse. The syndrome is made up of two separate, but related disorders: Wernicke's encephalopathy and Korsakoff's psychosis.

The syndrome is named after the two people who first described the conditions. Karl Wernicke, a Polish neurologist, published his description of Wernicke's encephalopathy in 1881 and Sergei Korsakoff, a Russian psychiatrist, published the first of a number of papers describing the condition which bears his name in 1887.

The most widely recognised characteristic of the syndrome is loss of memory. Wernicke's encephalopathy usually precedes Korsakoff's psychosis. The symptoms and likely consequences of the two conditions are different so this factsheet describes them separately.

Who develops it?

Post-mortem studies suggest that the Wernicke-Korsakoff syndrome occurs in about 2% of the general population and 12.5% of dependent drinkers (1). The vast majority of sufferers develop the syndrome as a result of malnutrition caused by heavy drinking over time, however it is important to note that it is also possible to develop Wernicke-Korsakoff syndrome without consuming alcohol, although this is rarer. Workers in alcohol services report that the syndrome affects people in their thirties, forties and fifties as well as those in older age groups. For an overview of how prolonged drinking affects the body, see factsheet 38, Damage to Health

Wernicke's encephalopathy

Wernicke's encephalopathy can develop quite suddenly and the symptoms are easy to miss or mistake for straightforward drunkenness. It has been reported that 17-20% (3) of people who develop Wernicke's encephalopathy die because of the condition and that 85% of survivors go on to develop Korsakoff's psychosis (3). Because the condition is difficult to spot, it is often only revealed by post-mortem examinations.

The symptoms of Wernicke's encephalopathy are:

- confusion about time and place
- drowsiness
- poor balance
- double vision
- abnormal eye movements or paralysis of eye muscles that might seem like a squint

Not all of the above symptoms will necessarily be present in every case and the person with Wernicke's encephalopathy may just appear to be confused for no good reason. If they are a known drinker or smell of alcohol it is easy to see how the condition is difficult to diagnose. If you know how much the person has drunk, a good clue the condition is developing can be that they appear more drunk than you would expect.

It is important that Wernicke's encephalopathy is diagnosed as soon as possible because it can be fatal and if it is left untreated irreparable brain damage may occur (this is the second phase of the syndrome; see below). A blood test can confirm thiamine deficiency, however this may take up valuable time and it is important that Wernicke's encephalopathy is treated with large doses of thiamine (vitamin B1) by intravenous or intramuscular injection as soon as possible.

Alcohol and brain damage

Heavy drinking over time is associated with a number of different types of brain damage (2) in addition to Wernicke-Korsakoff syndrome; these include:

- Cerebellar degeneration
- Alcoholic dementia
- Stroke
- Hepatic encephalopathy
- Marchiafava-Bignami disease
- Central pontine myelinolysis

Alcohol and thiamine deficiency

Heavy drinking can cause gastritis which is an inflammation of the stomach. If someone has gastritis they will often vomit up food before it can be digested. If food does not make it to the stomach, it will not be absorbed properly because the stomach is inflamed. Because it affects how much and how well food is absorbed gastritis can lead to malnutrition and therefore contributes to low levels of thiamine (vitamin B1) in problem drinkers. The liver is a complex organ with many vital roles. It can affect the body's ability to absorb and store nutrients such as thiamine. There are 3 stages of liver damage associated with heavy drinking:

- fatty liver
- alcoholic hepatitis
- alcoholic cirrhosis

What causes Wernicke's encephalopathy

Wernicke's encephalopathy is caused by a lack of thiamine (vitamin B1)

Many heavy drinkers don't eat properly because alcohol takes the place of food. When they do eat they can have difficulty absorbing nutrients because their stomach or liver may be damaged. This can lead to malnutrition and vitamin deficiencies.

The body uses thiamine to form part of the structure of blood vessels, including those in the brain. Without it they begin to leak. The main symptoms of Wernicke's encephalopathy (confusion, drowsiness etc) occur when blood is leaking out of the capillaries in and around the parts of the brain that form new memories and the parts that control muscle co-ordination. If the leaking continues the patient will probably become unconscious, the leaked blood then clots and as it solidifies damages the brain tissue around it.

Treatment of Wernicke's encephalopathy

Wernicke's encephalopathy can be treated by high-dose injections of thiamine (for dosage recommendations see ref.1). The patient who receives thiamine in this way can recover fully and be left with no memory problems, although this is not guaranteed. However, if the patient does not receive large doses of thiamine before becoming unconscious it is more likely that they will have sustained permanent damage and will begin to show signs of Korsakoff's psychosis. Signs of damage to the person's memory may become obvious in a matter of weeks, or it could take longer for the symptoms to become obvious.

Individuals who are malnourished or who have been drinking heavily will not be able to absorb thiamine (vitamin B1) very well through their digestive system. Therefore, it is important that people who have developed Wernicke's encephalopathy receive high-dose intramuscular or intravenous injections of thiamine rather than orally administered doses. This treatment can also prevent Wernicke's encephalopathy from developing. Unfortunately, there has been a trend away from prescribing this vital treatment. This is possibly due to a combination of factors including a warning in 1989 from the Committee on Safety of Medicines about possible serious side-effects from the only injectable vitamin B supplement then available. It seems likely that the committee have overemphasised the risks associated with injecting vitamin B and underestimated the risks and prevalence of Wernicke's encephalopathy (3). This may be leading to an increase in the number of cases of Wernicke-Korsakoff syndrome.

It is important to appreciate that people with Wernicke's encephalopathy may come into contact with doctors and health professionals in a wide variety of situations and for a range of reasons. For example someone who has burnt themselves whilst drunk may be admitted to a burns unit and the symptoms may not be identified as a separate condition. Because high-dose injections of thiamine can both prevent and successfully treat Wernicke's encephalopathy, it is vital that health professionals, and others who work with people who misuse alcohol, are able to identify the condition and the risk factors associated with it so that sufferers and people likely to develop it can be appropriately treated.

Half the people who go to hospital with a head injury are heavy drinkers (4), assessment of their condition should include consideration of the need to administer high-dose injections of thiamine. Places like accident and emergency departments need to develop clear and objective guidance about how to identify and deal with people who have Wernicke-Korsakoff syndrome or may be likely to develop it.

Injection for prevention and treatment

It has been suggested that all patients undergoing hospital-based detoxification and patients with a history of alcohol misuse who experience moderate to severe withdrawal-related neuropsychiatric symptoms should be treated with high-potency B-complex vitamins (2). The aim of this suggestion is to prevent the significant fatalities and ill effects associated with Wernicke-Korsakoff syndrome. Wernicke's encephalopathy is often not diagnosed (80-95% of cases) (2), so routine injections of high-level B complex vitamins would appear to be a logical way to improve the treatment and prevention of the syndrome. However, the idea of routine high-dose intra-muscular or intra-venous injections of B-complex vitamins for individuals undergoing detoxification becomes complicated as more and more people are detoxed in a home or community setting. The level of medical expertise and supervision required for a regular regime of high-dose B-complex injections is unlikely to be found outside of a hospital or residential setting. Until there is clearer guidance from the Committee on Safety of Medicines the matter will remain a point of debate and a decision for individual alcohol service managers and clinicians.

Korsakoff's psychosis

If Wernicke's encephalopathy is untreated Korsakoff's psychosis can develop, although confusingly it can also develop without an episode of Wernicke's encephalopathy. The symptoms of Korsakoff's syndrome are:

- loss of memory, particularly short term memory
- loss of spontaneity and initiative
- confabulation (presenting inaccurate accounts of personal history of recent events)

The symptoms can range from mild to severe.

The above symptoms are caused by damage to specific areas deep within the brain. This damage is caused by the effects of lack of thiamine and takes place during an episode of Wernicke's encephalopathy.

Because alcohol is a toxic substance which comes into direct contact with the brain (once absorbed into the blood stream), heavy drinking over time also leads to more general damage to the outer layers of the brain which can also affect memory and behaviour.

Korsakoff's psychosis is the term most commonly used to describe patients who have problems with their memory, but whose understanding and other mental processes are mainly unaffected. If the person's intellect and understanding is more generally affected then the term 'alcoholic dementia' may be used. There can be an overlap between these terms (see page 5 for more details.)

How does Korsakoff's affect the person?

The most obvious symptom of Korsakoff's syndrome is memory loss. Often it is recent memories that are affected, although the ability to recall the more distant past can also be affected.

Memory loss

Because the part of the brain responsible for creating new memories has been damaged a person with Korsakoff's psychosis may appear to forget the incidents of their daily life as soon as they occur. Korsakoff's patients may have memories from their earlier life, but know little about their more recent past. The technical term for this inability to remember new information for more than a few seconds is anterograde amnesia. If the patient has experienced an episode of Wernicke's encephalopathy then the date of that episode may be the cut-off point after which they are unable to retain memories for very long. They will however, be able to remember events from before the episode. This can lead to many practical difficulties: WKS sufferers may forget people or conversations moments after they have taken place or they may need weeks of practice to learn simple information. For example, if they move to new accommodation, they may need extra help or time to learn the route from their bedroom to the kitchen.

Thiamine (vitamin B1)

Thiamine can be found in beans, green vegetables, sweet corn, egg yolk, liver, cornmeal and brown rice. Thiamine is added to staples such as flour in the UK. As well as Wernicke-Korsakoff syndrome lack of thiamine can also cause beriberi. The main symptoms of this are fluid retention, heart disease and inflammation of the nerves. Heavy drinking is the commonest cause of thiamine (vitamin B1) deficiency in Western countries.

Two types of confabulation

Recent research (5) suggests that there are two types of confabulation:

Spontaneous confabulation which is the production of a wide range of false memories and fantasies without external prompting. Spontaneous confabulation occurs in patients with considerable frontal brain damage or advanced dementia.

Provoked confabulation which is the plausible but inaccurate recall of recent or long past events which have been presented out of their correct context in response to questioning. Provoked confabulation is more usual of the kind of distortion of reminiscence found in Korsakoff sufferers.

This research suggests that the confabulation exhibited by Korsakoff patients is a normal response to a badly damaged memory system.

How does Korsakoff's affect the person? (continued)

Confabulation

Some Korsakoff sufferers may fill in these gaps in their memory with made-up memories or fantasies, a process known as confabulation. Our brains often fill in gaps when information is missing or confusing (e.g. when faced with an optical illusion) and confabulation is this normal process, but on a grander scale. The gap in memory may be filled with a set of events constructed around the few fragments of real memory that remain. Patients may transplant events that they can remember from their earlier life and apply them to their current circumstances. Although inaccurate, the events are fully believed by the sufferer.

Occasionally patients develop what may appear to be delusions about their past. A sufferer might consistently explain their current situation, for example of being cared for in a home, by stating that they were in the armed forces and are now convalescing following active service. Examples of confabulation to this degree are extremely unusual, but they give a clear illustration of how confabulation is a subconscious attempt by the sufferer to make sense of their situation when recent memories and information are missing.

Overall, confabulation is experienced by a small number of Korsakoff sufferers and most Korsakoff patients respond to questions about their recent past by acknowledging that they cannot remember.

Other impairments

Most Korsakoff sufferers are aware of their surroundings and will understand, if they are in hospital or living in care, where they are. They will eventually be able to learn their way around their environment, but will remain unable to gauge the passing of time and will be unclear about how long ago things have happened.

Although overall intelligence can remain intact there is often a deterioration in the sufferer's ability to think or understand (cognitive impairment). Korsakoff's sufferers may have impaired attention and difficulty in 'screening out' irrelevant information. They are thus likely to be easily distracted or lose concentration. For example, sufferers, may have more difficulty completing fragmented picture puzzles or in telling whether two colours, smells or songs are the same or different.

A further symptom of Korsakoff's syndrome is a loss of spontaneity and initiative. In conversation sufferers usually respond to questions straightforwardly with little elaboration, their answers can seem brief or superficial and they are generally not bothered about keeping conversations going. If left to themselves, groups of Korsakoff patients rapidly sink into silence, with no sign that this causes them any distress (6). There are exceptions to this, some sufferers are chatty, but they will usually repeat the same stories or concerns over and over again.

Most people with amnesia become distressed when presented with evidence of their disability, however people with Korsakoff's tend to lack insight into the significance of their condition or even how they might have developed it. Most patients are unconcerned even when faced with clear evidence of their memory's failure. This is a characteristic shared with people suffering from dementia such as Alzheimer's disease.

Because there is a specific point at which people's memory has been damaged, many of the skills learnt before this 'cut-off point' will not be affected. This will usually include things like speech, language and gesture and sufferers may have no problems with basic activities of daily living unless they are moved to a new living environment which would require them to memorise or process new information.

General effects of alcohol-induced brain damage.

Heavy drinking over time can have serious repercussions for the functioning of the entire nervous system, but particularly the brain. These effects include:

- Poor attention
- The need for extra time to process visual information
- Self-centred and withdrawn behaviour
- Lack of sensitivity in social situations
- Difficulty with visuospatial abilities (dealing with objects in 2 or 3 dimensional space)
- Difficulty with problem-solving and learning new information
- Mild to moderate impairment in short and long term memory.

The condition of generalised alcohol-related brain damage which affects behaviour is given the name of 'alcoholic dementia'. Even if general intelligence appears intact, brain scans show loss of brain tissue and brain weight is significantly reduced in heavy drinkers at death. Abstinence can lead to improvements, possibly more so in women and younger people.

Wernicke-Korsakoff's syndrome, dementia and other brain disorders: where to draw the line?

The symptoms and development of Wernicke-Korsakoff's syndrome can vary from patient to patient. Some people may be more severely affected than others. The evolution of problems with memory, understanding and behaviour varies among sufferers and may depend on how severely or frequently the person has experienced thiamine deficiency.

Because people who develop the syndrome are likely to have been drinking at high levels, they may have sustained other alcohol-related brain damage. Research is not yet able to show what amount of alcohol intake leads to what type of physical damage within the brain.

The most popular view is that the specifically located brain damage that causes Wernicke-Korsakoff's syndrome occurs in addition to the more widely-distributed brain damage caused by dependant drinking. Thus sufferers may experience a steady decline in their abilities (due to widely-distributed brain damage) which may change to the more dramatic amnesia of Korsakoff's syndrome after an episode of Wernicke's encephalopathy. The more generalised brain damage will also be present.

The line between what is Wernicke-Korsakoff's syndrome, or alcoholic dementia or milder brain damage is thus quite a hazy one. Diagnosis and the decision about what name a sufferer's condition is given depends on the sufferer's behaviour and how much can be found out about the physical damage. It is hard to work out which bit of the brain is damaged and if it is, exactly how it effects their behaviour. Technical developments such as brain scans are improving our ability to relate physical damage to particular kinds of behaviour.

People who could be diagnosed as having Wernicke-Korsakoff syndrome will have a range of symptoms which will vary in their severity and probably be complicated by a variety of medical, social and mental health problems as well as vastly differing histories of alcohol use.

Are the effects of Wernicke-Korsakoff syndrome permanent?

There has not been very much research carried out about the rehabilitation of patients with alcoholic dementia or Korsakoff's syndrome. However there is research that shows that problems with memory, awareness and understanding can improve, to various degrees, in many sufferers with time. The physical or chemical processes that lead to this improvement are not understood, but studies of Korsakoff's sufferers and an animal model of Wernicke-Korsakoff syndrome suggest that undamaged parts of the brain that deal with memory, awareness and understanding can compensate for the damaged areas and lost abilities. This compensatory ability of the brain is huge at birth but declines with age. Drinking alcohol, smoking, head injury and severe infections may further reduce the brain's reserve (7).

While the amnesia itself may not respond to treatment, stopping drinking and an intensive programme of vitamin therapy followed by tailored therapeutic rehabilitation can lead to significant improvements. Some sufferers may be able to live independently, however others will need long-term residential care or structured support in the community.

'What is this?' I asked showing him a photo in the magazine I was holding. 'It's the moon' he replied. 'No it's not' I answered. 'It's a picture of the earth taken from the moon' 'Doc, you're kidding! Someone would've had to get a camera up there!' 'Naturally' 'Hell! You're joking – how the hell would you do that?' Unless they were a consummate actor, a fraud simulating astonishment he did not feel, this was an utterly convincing demonstration that he was still in the past. His words, his feelings, his innocent wonder, his struggle to make sense of what he saw were precisely those of an intelligent young man in the forties...

'He is, as it were,' I wrote in my notes, 'isolated in a single moment of being, with a moat or lacuna of forgetting all round him. He is a man without a past (or future), stuck in a constantly changing, meaningless moment' And then, more prosaically, 'The remainder of the neurological examination is normal. Impression: probably Korsakoff's syndrome, due to alcoholic degeneration of the mammillary bodies'

Oliver Sachs in 'The man who mistook his wife for a hat' London: Picador (1985)

Treatment for people with Korsakoff's psychosis

It is important to note that Korsakoff's psychosis is not inherently progressive in the same way that an 'organic' cause of dementia such as Alzheimer's disease is. Overall, Wernicke-Korsakoff syndrome should be viewed as a treatable condition. The question of how successful treatment of someone with Korsakoff's psychosis will be depends on the degree of damage done to the brain.

People who have developed Wernicke-Korsakoff syndrome are likely to have been drinking at high levels for a number of years. This will probably have affected their relationships with family and friends and they are unlikely to have good relations or close contact with many people. It also means that they may be affected by other disorders related to alcohol misuse e.g. cirrhosis, pancreatitis etc. Their treatment needs are therefore likely to be complex and long-term.

Assessment of the individual should focus on identifying the degree of damage sustained. How intact the person is in terms of their social functioning and understanding can then be translated into an assessment of what skills they are missing. Treatment should then either focus on changing the patient's limitations (practical, social and memory skills) or by changing the environment in which they live.

If it is suspected that someone has Wernicke-Korsakoff syndrome and they are still drinking, they will need emergency in-patient care so that the necessary detoxification and initial vitamin treatment can take place. Further assessments can be made after the sufferer has stopped drinking and successfully withdrawn from being dependent on alcohol (detoxification).

In addition to general observation and routine mental state examination, a full assessment of the sufferer's condition will involve skilled psychological testing and brain imaging. These two assessment approaches are complementary: testing and scans can confirm the presence of significant impairment and damage; repeated assessment will indicate what level of recovery (if any) is likely. It is important to note that testing carried out immediately after detoxification can give misleadingly pessimistic results. Many of the problems that individuals may have with their behaviour and ability to understand can improve after detoxification, others seem to require practice for improvements to be made. Studies show that older and more severely impaired sufferers do not recover as well as others once they stop drinking.

It will be possible to start on therapeutic work to improve the sufferer's practical and social skills once the person with Korsakoff's psychosis is orientated and settled in their surroundings. Although sufferers may not be upset by their loss of ability to remember, moving into a new environment or not being familiar with rules or routine because they are unable to remember may cause them stress and anxiety. The first goal of treatment or care should be to establish physical and mental stability and to reduce any excessive stresses felt by the individual. Once the sufferer feels secure that their basic needs such as food, warmth and clothing will be met it will be possible to start to address their lost social and practical skills.

Therapeutic work with people who have memory disorders tends to focus on the effect of their lack of memory on their behaviour. The aim is to work out practical solutions when their lack of memory skills will affect their everyday lives, for example learning to write down the names of friends.

Memory rehabilitation

There are a number of therapeutic techniques that can help people who have memory problems. They can include things like learning by rote or responding to visual imagery or using word association.

The aim of memory rehabilitation will fall into one of the following categories:

- To improve or retrain memory skills
- To teach people ways to bypass their memory deficits as far as possible
- To discover the most efficient way for individuals to retain new information.

Whatever method or theory is employed, other factors such as living arrangements and family and social support will play an important part in an individual's rehabilitation. Problems may arise for Korsakoff sufferers when potential solutions such as 'face-name mnemonic imagery techniques' which can aid the ability to remember people's names, are difficult to apply and need quite a lot of motivation to learn in the first place.

Helpful texts

Edwards, G. et al (1997), *The treatment of drinking problems : a guide for the helping professions* (3rd ed, Cambridge : Cambridge University Press.)

Ward, M (1998) *Caring for someone with an alcohol problem.* (London: Age Concern.)

Treatment for people with Korsakoff's psychosis (continued)

Along with therapeutic work that focuses on the sufferer's memory and behaviour it may also be necessary to address their alcohol problem, although for some people this ceases to be an issue as they effectively forget their need to drink and just need reminding not to drink. For other sufferers who may be experiencing difficulty with their awareness and understanding, but still want to drink, a traditional, demanding, therapeutic alcohol treatment regime will be inappropriate. Rather than expecting the Korsakoff's sufferer to join in with an established alcohol counselling programme, it will be necessary to design a programme around their needs. 'the best therapeutic programmes involve a great deal of new learning, emotional awareness, and social interaction, and it is exactly the skills needed to engage successfully in those types of experience in which the brain-damaged person may be deficient' (8).

Sources

1. **Cook, C.H.C & Thompson, A.D** (1997), B-complex vitamins in the prophylaxis and treatment of Wernicke-Korsakoff syndrome. *British Journal of Hospital Medicine*, vol.57, no.9, pp 461-465
2. **Lehman, L.B. et al** (1993), Neurological disorders resulting from alcoholism, *Alcohol Health and Research World*, vol.17, no.4, pp305-309
3. **Thomson, A.D & Cook, C.H** (1997), Parenteral thiamine and Wernicke's encephalopathy: the balance of risks and perception of concern. *Alcohol and Alcoholism*, vol.32, no.3, pp207-209
4. **Ferguson, R.K. et al** (1997), Thiamine deficiency in head injury: a missed insult? *Alcohol and Alcoholism*, vol.32, no.4, pp493-500.
5. **Kopelman, M.D** (1987), Two types of confabulation. *Journal of Neurology, Neurosurgery and Psychiatry*, vol.50, pp1482-1487.
6. **Knight, R.G & Longmore, B.E** (1994), *Clinical neuropsychology of Alcoholism.* Hove: Lawrence Erlbaum Associates, p269.
7. **Zeman, S** (1997), *All about dementia.* London: The Mental Health Foundation.
8. **Edwards, G. et al** (1997), *The treatment of drinking problems : a guide for the helping professions* (3rd ed, Cambridge : Cambridge University Press.)

Addresses

Carers National Association

20-25 Glasshouse Yard
London, EC1A 4JS
Tel: 020 7490 8818
Helpline (1-4pm) – 020 7490 8898

Alzheimer's Disease Society

Gordon House, 10 Greatcoat Place
London, SW1P 1PH
Tel: 020 7306 0606

Mental Health Foundation

20-21 Cornwell Terrace
London, NW1 4QL
Tel: 020 7535 7400

Medical Council on Alcoholism

3 St Andrews Place
London, NW1 4LB
Tel: 020 7487 4445

Services

The following organisations offer residential care for people affected by Wernicke-Korsakoff syndrome. Alcohol Concern would welcome details of other facilities who offer services to this client group. Inclusion in this list does not indicate endorsement by Alcohol Concern

Chaucer Clinic

St Bernards Hospital Estate
Uxbridge Road
Southall, Middlesex,
UB2 4XB
Tel: 020 8571 4616
Fax: 020 8571 0087

Dyfrig House

South Glamorgan Council on Alcohol
53a Fitzhamon Embankment
Riverside, Cardiff, CF1 8RN
Tel: 01222 232 127
Fax: 01222 226 660

Farmstead Nursing Home

New Road
Hellingly, Hailsham
East Sussex, BN27 4EW
Tel: 01323 442050

Heavy Drinkers Project

Manchester Methodist Housing Association
450 Great Western Street
Manchester, M14 4NA
Tel: 0161 248 9069
Fax: 0161 225 3242

Leadbeater Homes

PO BOX 76
Margate
Kent, CT9 2YJ
Tel: 01843 220 715
Fax: 01843 231 639

Tapton Grove Continuing Care Unit

Grove Road, Brimington
Chesterfield, Derbyshire
S41 1QH
Tel: 01246 274 178

Guys Cross Nursing Home

122 Coventry Road
Warwick, CV34 5HL
Tel: 01926 773920

The Jasmine Centre

4 Canterbury Road
Westgate on Sea
Kent, CT8 8JJ
Tel: 01843 231677
Fax: 01843 231639