WERNICKE'S ENCEPHALOPATHY: LOST AND FOUND

Translation of the case history section of the original manuscript by Carl Wernicke "Lehrbuch der Gehirnkrankheiten fur Aerte and Studirende" (1881) with a commentary

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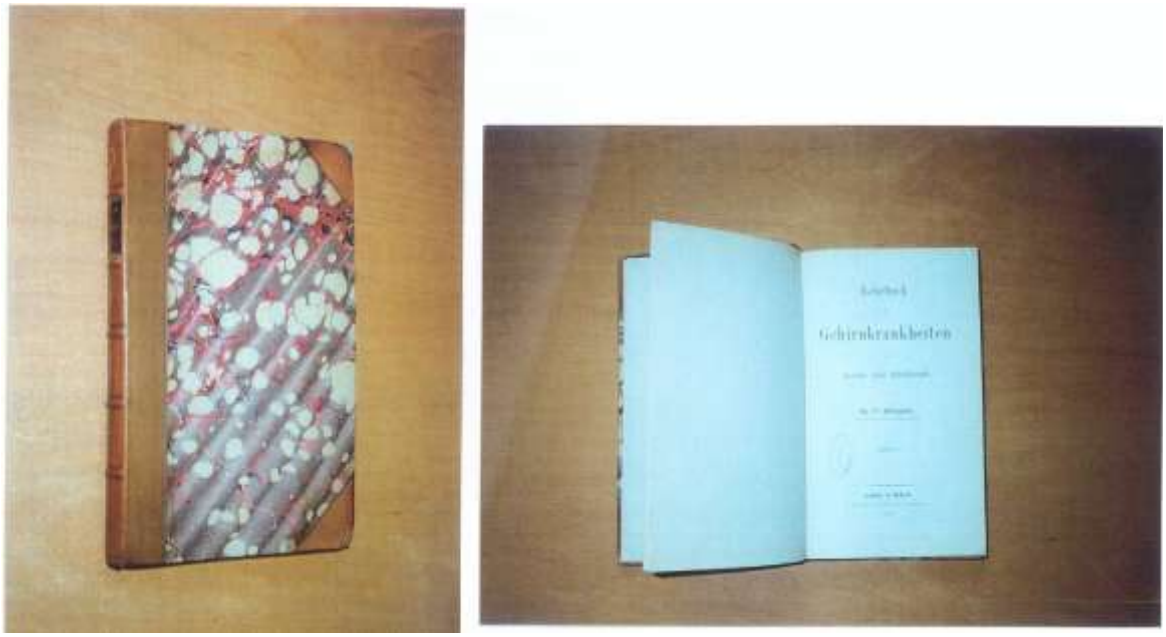
Abstract-Aim: A translation into English of the case history section of Carl Wernicke's original manuscript of 1881 with a discussion on its relevance for clinicians of today.

It is 125 years since Carl Wernicke published his *Lehrbuch der Gehirnkrankheiten fur Aerte and Studirende* in 1881 (Figure 1) in which he described a new disease, based on clinical and pathological observations in three patients (Wernicke, 1881). It is also 100 years since the first description of Alzheimer's disease (Alzheimer, 1904). Both of these diseases cause devastating neurological sequelae and neither has specific ante-mortem diagnostic tests. Recent publications estimate that approximately 24.3 million people in the world have some form of dementia (Ferri et al, 2005), indicating an urgent need, where possible, to identify the cause of the dementia and to institute preventative or early and effective treatment before irreversible damage occurs.

FIGURE 1. Wernicke, C. (1881). Lehrbuch der Gehirnkrankheiten fur Aerzte und Studirende. Vol 2 Kassel Theodor Fischer 229-242



The concept of a "classic triad" of signs and symptoms in Wernicke's Encephaopathy (WE) was based on the original description by Wernicke. The triad consists of an abrupt onset of a confusional state and impairment of

consciousness, ataxia and eye signs (nystagmus and ophthalmoplegia). Similarly, Victor et al (1989) in their famous monograph concluded: *"The diagnosis of Wernicke's disease is made most readily on the basis of the acute appearance of ocular palsies, nystagmus, ataxia of gait, and disturbances of consciousness and mentation, which may present singly or in various combinations. More than 80% of patients show signs of polyneuropathy as well, and associated liver disease is found in two thirds of the patients"* (Victor et al, 1989). However, as Wernicke himself described, and has been shown in subsequent studies, other important clinical signs and symptoms are often present before the later "classical" signs appear.

In reviewing the literature (DeWardener & Lennox, 1947; Cravioto et al., 1961; Torvik et al., 1982; Harper et al., 1986; Lindboe & Loberg, 1989; Fattal-Valevski et al., 2005), we have been struck by the consistency of the reported signs and symptoms attributed to WE. Despite this, clinicians continue to focus on the triad, almost in a perseverative manner! In exploring why this is, we have taken an historical/investigative perspective on the situation and returned to the primary literature sources. The original detailed descriptions of Wernicke's patients would seem a good place to begin. In 1989 Victor and his colleagues reviewed the three cases described by Wernicke, together with many published case reports from the early 20th century (Victor et al, 1989). We are not aware of any English translation of Wernicke's original German paper so this was organized by one of us (CCHC) and is presented below. Wernicke described the early signs and symptoms of his eponymous encephalopathy in three untreated patients.

WERNICKE'S CLINICAL DESCRIPTIONS

> A 20-year old seamstress, was admitted to the Charité on *5 December 1876* with sulphuric acid poisoning, and was discharged, cured, on *6 January 1877*. She said that she had vomited very soon after leaving the institution but had otherwise been well until *3 February 1877*, after which she was confined to her bed and was noticeably sleepy, yawned a good deal and staggered as she walked whenever she attempted to leave her bed. The patient also noticed a deterioration in her eyesight, and later also a constant, very irritating flickering and a strong dislike of light, as well as giddiness and heavy-headedness. Since all these complaints increased and the vomiting persisted, she had herself admitted to the Charité on *11 February 1877*. There had never been a specific infection. On *12 February* her condition was as follows: The patient, very pale, and showing some loss of weight, is lying with her eyes half closed. She opens the lids [/ palpebral fissures] only about 1 cm and cannot open them further even if her eyes are shaded. When looking upwards she opens the eyes a little wider. The opening of the right eye [palpebral fissure] **[234]** is normally, and also when looking upwards, somewhat narrower than that of the left. The eyes close completely but with little energy, without the formation of a fold, nor can the eye be induced to close more firmly as a reflex response

to being touched. When looking fixedly straight ahead the two eyeballs are stationary and slightly convergent. When looking upwards, spasmodic movements occur in large excursions, but in the end the movement succeeds to a great extent. The same is observable when the patient looks downwards. The movement of both eyes to the left is significantly impaired. When doing this the left eye will not, despite visible effort and spasmodic movements, go beyond the mid-line, while the right eye can move further inwards, but the inner edge of the cornea can be brought close to the caruncle only for a few moments; generally the inner edge of the cornea only reaches the line of the lower lacrimal point. Movements to the right are similarly impaired: the right eye can also only be brought to the mid-line, while the left can move better than the right to the inner corner, but is still noticeably restricted. When the visual axes are made to converge (by fixing on the point of the nose), the same behaviour is apparent; the left eye is better at turning inwards than the right. When at rest, the aberration of the ocular axes is not very noticeable, although there is clear convergence. The pupils, which are equal and at medium dilation, react sluggishly to light. When the face is at rest the right-hand corner of the mouth is lower and the right cheek is somewhat affected [abnormally smooth?], while on the left side the nasolabial fold and also the fold marking the edge of the lower eyelid are strongly visible. The right corner of the mouth is less open than the left. The expression is as though on the verge of tears, stubborn, and at the same time apathetic. When the patient laughs or opens her mouth wide the same difference remains, whereas when other voluntary movements are made no difference is perceptible between the two halves of the face. There is no other evidence of paralysis, but there is a great feeling of weakness. Patient can walk with support. No loss of sensation; the extremities are cold. Patient complains of extreme tiredness, and her speech is also tired and as though she had just woken from sleep. Repeated vomiting in the course of the day. T. 37.0 -- 37.4. *13 February.* Somnolence persists, patient merely emits a groan from time to time or calls out the name of her fiancé, she is altogether disorientated and seems not to know where she is; when she is wide awake, e.g. at meals, she talks rationally with the staff but suddenly interrupts the conversation, complaining of pain in the small of her back and heavy-headedness. Urine passed sparingly, of a peculiarly oily consistency, contains peptone, no protein, no sugar. Cool extremities. T. 36.4 -- 37.2. *14 February.* Patient has to be woken up, answers but remains disorientated. Complains of headache and stiff neck. Shows extreme anxiety **[235]** and fear of falling when she is about to be carried. Frequent yawning and groaning. She answers only when questioned repeatedly. Pupils dilated by atropine. Ophthalmoscopically, neuritis optici in both eyes, with only moderate swelling and many streaks of blood. Abdomen bloated, very tight and painful. T. 37.3--37.5. P. 120, barely perceptible. Very restless during the night, often crying out. *15 February.* Soporous state, whimpering with pain, does not answer when spoken to. Death during the afternoon.

Autopsy. There is no flattening of the gyri, the cerebral ventricles contain only a few drops of fluid, tela and plexus are bright red. On section of the central ganglia one sees, along the full length of the third ventricle, in its walls, for a distance of about 3--5 mm, an overall pink coloration of the adjacent cerebral mass, in which numerous small, punctate haemorrhages are present. The lesions are, with almost mathematical precision, equal on both sides. The section shows them very clearly in the area of the middle commissure, which is well developed and is also permeated with haemorrhages; no similar lesions can be observed any longer in the quadrigeminal bodies, nor is there anything in the cerebellum. The pia on the medulla oblongata has a smoke-grey colouring; in the pons and med. oblongata there are no lesions of any size. In the spinal cord no macroscopically visible lesion is present. Numerous haemorrhages in both retinae. Apart from that, traces of the sulphuric acid poisoning. Anatomical diagnosis: encephalitis haemorrhagica substantiae griseae ventricul. III. Haemorrhagiae retinae utriusque. Stenosis pylorica ventriculi; Gastrectasia intorixatione sulfurica, chronica, ulcerosa. The examination made after hardening revealed that the haemorrhages mostly enclosed [/ invaginated?] the vessels, the punctate ones varied in size and in isolated cases attained the size of a pinhead. The small vessels and capillaries much distended and very full, the vessel wall showing no noticeable lesions, only here and there there appeared to be swelling of the capillaries and endothelial cells of unusual size. Close to the haemorrhages there were always granular cells. As for the distribution of these lesions, they never reached the formations and fibre masses adjacent to the grey floor. Only in the posterior quadrigeminal ganglion on the left-hand side was there, right at the centre, an isolated, pinhead-sized haemorrhage. Going downwards this same lesion in the grey

floor reached, with gradually decreasing intensity, to the area of the striae acusticae. The basal arteries appear normal.

A 36-year-old Scot, a piano teacher and allegedly a professor, is a heavy drinker and has drunk cognac, in particular, in large quantities. **[236]** He is admitted to the Charité in a delirious state on *18 June 1877*, has a sleepless night, continuously delirious but not noisy. Condition on *19 June* as follows: a well-nourished, somewhat corpulent man with no disease of the inner organs, talking in delirium in a low voice, shows great motor restlessness [/ superactivity] and very strong tremor, is utterly drenched in sweat. Taken from his bed, patient can walk only very unsteadily and with support, his gait is atactic, interrupted by jerky, purposeless movements, with legs wide apart and trunk held rigid. The patient's restlessness precludes further examination of him lying on the bed, except that it is noted that he responds to pin-pricks to the sole of his foot. His hands move freely, their movement impaired only by the tremor. His speech also extremely tremulous; difficult to assess it, as patient speaks Scottish and only mutters disjointed sounds to himself, and it is impossible to capture and hold his attention. Both pupils very much contracted, almost to the size of pinheads; both the same. The mobility of the eyes seems to be impaired, but it is impossible to assess this more precisely as patient does not focus. Skin temperature not raised. Respiration accelerated, pulse also accelerated, fairly weak, regular. In the following days the delirium continued unchanged, the patient's general condition showed no cause for anxiety. It was seen in the course of repeated observation that the patient barely moved his eyes at all, and was able to focus on objects only by moving his head. With the decline in restlessness there began a somnolent state combined with extreme weakness; patient was unable to maintain a standing position. The pulse, always very soft, became ever weaker, faster and also irregular, the respiration too became irregular and patient died after a fairly long sopor on *26 June*. In his calm period it was possible to test more effectively the mobility of his eyes, and it could be seen that the eyes were almost wholly fixed in the middle position, but there was no ptosis. The pupils, which were otherwise narrowed, dilated with the use of atropine. Ophthalmoscopically the papilla of each eye appeared noticeably reddened, but without any swelling, and in the right eye there was a linear haemorrhage along one vessel.

Autopsy. In the brain the same findings as in the previous case. The haemorrhages are nowhere more than punctate and are confined to the central cavity grey [matter] of the third and fourth ventricle and the aqueduct. The area of the ganglion hanenulae is also affected on both sides. Microscopically the lesions are exactly like those in the previous case. Basal arteries normal.

33-year-old man is admitted to the Charité on *10 March 1878* after the onset of delirium tremens in the morning. A heavy schnapps drinker, especially in the previous few months. Since the last campaign **[237]** frequent complaints of aches in his legs, on an earlier campaign had typhus, once infected as a young man. In the last 4 weeks his walking had been poor at times. 3 weeks ago he had difficulty in passing water, lasting for a week. 8 weeks ago patient is said to have once fallen from a wagon, perhaps on to his head, but without any resultant symptoms. In the last 4 weeks giddiness and headaches at times, and also vomiting in the mornings, especially after heavy drinking. In the past 2 days patient has complained of double vision, and has had jaundice for about the same length of time. Has never had a stroke or attacks of cramp. On *11 March* condition as follows: A very strong, well-nourished individual with slightly icteric colouring, an old scar from a bubo in the right inguinal area. On the transitional fold of the penis a stringy, shiny, scar-like cord; on the lower legs several circular, clearly-outlined patches lacking pigmentation. Since his admission patient has behaved wholly like a man in delirium, is disorientated as to place and surroundings. Now very considerable motor restlessness [/ superactivity], his hands constantly pluck at the blanket, patient throws his body about and turns his head to focus on people who are not there, he calls out the names of acquaintances and also imagines frightening things. As he gesticulates energetically, a strong tremor is present. Voice hoarse, speech hasty, words falling over each other, somewhat tremulous, not incoherent, and no anarthria. Face covered with sweat, respiration much accelerated, apparently as a result of mental agitation, no cough. Complexion slightly cyanotic, bloated appearance. The lips and teeth have a crusted, bark-like coating. A staggering gait, also impaired by sudden atactical movements and tremors. Is

able to stand for a short time on either leg. When asked, patient complains of weakness in his legs. Stretched posture of trunk and head. Tongue somewhat dry, without traces of biting, is stretched out straight but with a strong tremor. In the face, very slight facial paresis on right-hand side. Patient is not confused, his attention can be held for some moments by questioning. P. 110--120, even, fairly full, very soft, T. 39.3. Urine free of protein and sugar. Organs of the chest normal, hepatic dullness extends beyond the costal arch by the width of two fingers. There is total paralysis of the abducens on both sides. Neither eye can be moved outwards beyond the mid-line. The other eye movements seem intact. Pupils of both eyes react to light and convergence of the axes of vision, are half-dilated and show no inequality. There is a prompt reaction to being pricked with a needle in the face and on the hands, but only slight reaction when pricked on the feet. Patient feels a deep prick with a needle in his right big toe only after some time, and then complains of a very strong burning sensation; in the left [toe / foot] most pricks produce no reaction. Tapping on the head not painful, nor is pressure on the spine and the neck muscles. In the evening **[238]** intense reddening of the papillae, but no swelling, was observed ophthalmoscopically; the edges of the papillae not quite sharp. No haemorrhages. Further increase in motor restlessness [/ superactivity] and sweating. P. 120, T. 39, resp. 28--30. *12 March.* In the night the patient was delirious, [muttering] quietly to himself, somewhat calmer towards the morning. Patient is now lying peacefully with eyes closed, but when examined immediately wakes up and is delirious, [muttering] quietly to himself. No jactitation any more. Resp. 24--30, P. 100, regular, still very soft, T. 38,9. Sometimes bursts of coughing with catarrhal sputum. Yesterday an enema with vinegar added produced a copious, mushy stool. Neck is still held rather stretched, especially when the patient is raising himself into a sitting position. Pressure on the processus spinosi of the upper dorsal vertebrae and on the neck muscles produces a strong facial grimace, but stroking the skin of the back does not have this effect. The reduced sensitivity of the lower extremities has given way to hyperaesthesia, which is especially notable in relation to the patient's confusion and shows itself particularly in response to pressure on the calf muscles on either leg. Paralysis of the eyes unchanged, tongue trembles more when stretched out, facialis as yesterday. Pupils dilated with atropine. Evening T. 38.7, P. 90. *13 March.* P. 114. Resp. 26. T. 37.9. Patient is coughing less, still talking to himself, is delirious and disorientated. Gives a start when touched. Hyperaesthesia of the muscles no longer present. With support, patient can walk a few steps, though unsteadily, walks with legs wide apart and stiffly. Eyes and face unchanged. In the evening P. 100--110. T. 37.8. *14 March.* P. 98--100. T. 37.8. Pulse small and very soft, but regular. Patient was delirious for most of the night but this morning is less confused than yesterday and is beginning to orientate himself. As patient is focusing well, his eye movements are more closely examined. The outward mobility is as before, today inward mobility also seems to be impaired, upward and downward mobility intact. When sitting up, definite stiffness of the neck. In the evening P. 100, T. 38.0. Urine free of protein and sugar, strong colour. *15 March.* During the night the patient was delirious at times, mostly quiet. Now patient is in sopor, from time to time there are twitching movements, now of an arm or a leg, now of the whole body. Pulse very small, soft, regular, c. 96, about half an hour ago it was barely perceptible. Respiration is snoring, irregular, and there are quite long pauses, followed first by weak, then by increasingly deep inward breaths. This lacks the regularity of the sequence in [Cheyne-]Stokes respiration. Pressure on either big toe results in the leg being energetically drawn back, and in the case of the right one also facial grimace. Still starts when touched unexpectedly. **[239]** The patellar reflex can be investigated in the left [leg] and is not present. Patient can be awakened by calling to him, and he then says that he is fine. Strong thirst, dry lips, no great impairment of speech. When rising into a sitting position still has stiffness of the neck and facial grimace. *Evening*: P. 100, T. 38.2. *16 March.* P. 96, beginning to become somewhat irregular, T. 38.5. General state little changed, patient still delirious at times but is on the whole quieter, the weakness has increased, patient mostly lies with his eyes half closed, only the whites visible. Frequent coughing, no expectoration. Taken from his bed the patient also shows greater weakness and his legs give way after a number of unsteady, stiff paces. When one calls to him the patient is fairly conscious for some moments, is orientated and makes an effort to focus on objects held in front of him. Complains of great weakness. The mobility of the eyes has declined further, in both eyes the inward movement is very limited and is accompanied by twitching movements, the movement downwards is also considerably reduced, upward movement is the best preserved but also appears somewhat limited.

Ophthalmoscopic finding: the right eye clearly presents the signs of neuritis optici. P. intensely reddened, perhaps somewhat swollen, the edges unclear. The arteries not visible, the vein very blocked and all its branches filled. Close to the p. a small vessel pointing towards the macula is enclosed by a spindle-shaped haemorrhage. No other haemorrhages. In his left eye the patient is only hyperaemic, the edges somewhat unclear, arteries and vein very full, right to the very smallest branches. Towards midday death occurred.

Autopsy. Pia equally transparent on the convexity and base, very slightly oedematous. Dura not stretched, no lesions, pia also shows no lesions. The hemispheres themselves show nothing significant, only a slight reddening at the cortex. No lesions on the base. Upon removal of the roof of the 3rd ventricle one can see that the substance of the middle commissure is permeated with small punctate haemorrhages. After removal of the cerebellum, the greyest parts of the grey floor are also sprinkled with tiny red dots; possibly also containing capillary haemorrhages. Otherwise nothing noteworthy. Bone marrow without abnormality. Heart somewhat enlarged, walls not hypertrophic. On the side of the mitral valve facing the atrium, several rather old, very coarse, knotted thickenings. Lungs both of considerable volume, oedematous, containing much blood. Organs of the neck very cyanotic, mucous membrane of the larynx thickened, superficial ulcerations on both vocal cords. Spleen broad, cake-shaped, also somewhat enlarged in thickness. Kidneys both very hyperaemic. Anatomical diagnosis: haemorrhagiae punctiformes multipl. commissurae mediae, **[240]** ventriculi tertii et substantiae griseae ventricul. quarti, Oedema et hyperaemia pulmonum. Endocarditis chronica mitralis. Hyperplasia lienis. Infiltratio adiposa hepatis, Ulcera ligg. vocal. The examination of the hardened preparation revealed exactly the same lesion as in the two cases already described, except that the haemorrhages nowhere reached the size of a pinhead. The lesion, which on the basis of the [autopsy] findings is to be interpreted as inflammatory, nowhere extended beyond the grey floor of the fourth ventricle, in the third ventricle it was not so pronounced as in the previous case, but it extended somewhat further downwards into the uppermost area of the calamus scriptorius.

What is present is an independent, inflammatory, acute nuclear disease in the area of the nerves of the eye muscles, leading to death in a period of 10--14 days. The focal symptoms consist of associated paralyses of the eye muscles, which rapidly arise, progress and finally lead to almost total paralysis of the eye muscles; but even then with the exception of certain muscles, such as the sphincter iridis or levator palpebrarum. The patient begins to stagger when walking and his gait shows a combination of stiffness and ataxia, which is most reminiscent of the ataxia of alcoholics. The general signs are very striking and consist of alterations of consciousness, either somnolence from the outset or a final phase of somnolence preceded by a longer phase of agitation. In addition, in all three cases the visual nerves were affected, with inflammatory lesions of the papillae. In every case the onset of the disease had been preceded by taking harmful substances, in the one case sulphuric acid poisoning, which had healed [leaving?] pyloric stenosis, in the other two cases an uncommonly high level of alcohol abuse. The question of whether in these latter cases the signs of delirium potatorum should be regarded as a complication or as a separate general manifestation pertaining to this disease may be posed but cannot be decided; at any rate what was present was not an instance of common delirium tremens but, at the least, delirium tremens complicated by the symptoms of poliencephalitis. However, it should be pointed out that the characteristic disorientation was also observed in the first case, the features of which were totally different from those of delirium [tremens]. Whereas the first observation **[241]** had not been at all understood while the patient was still alive, in the second and third cases I was able to make the diagnosis despite the difference in the general signs. I think that this provides a justification for drawing attention to these cases as [representing] a particular syndrome.

Only one analogous case is to be found in the literature, and I only learned of it later. The similarity in the main symptoms and in the autopsy findings is unmistakable, even though it was not, as in our cases, only the nuclear region that was affected. The course [of the disease], lasting for five months, corresponds more to a sub-acute form of the disease.

Gayet. Affection encéphalique (encéphalite diffuse probable) localisée aux étages supérieurs des pédoncoules cérébraux et aux couches optiques, ainsi qu'au plancher du quatrième ventricule et aux parois

latérales du troisième. Arch. de phys. 1875. 28-year-old man, is admitted on *23 November*. In the middle of September he had been working in an engine-room when suddenly a boiler exploded not far from him. Although this caused great damage, he sustained no injury at all, but he became quite beside himself, could not sleep and was extremely agitated. He was able to work for three more days, but then he discovered that he could no longer read and write properly. After that he remained in a state of general weakness, exhaustion and apathy, and in addition, after a time, constant sleepiness. On admission he is able to answer perfectly intelligently, but slowly, and has to be kept alert. His facial features are mask-like, the orbicularis oris and palpebrarum are pressed close to the bones beneath them, there is ptosis on both sides and the lids [/ palpebral fissures] are 3/4 closed. Despite the very noticeable atonia there is no actual facial paralysis, all movements can be performed, but speaking, for instance, takes place as it were in isolation, without participation by the rest of the physiognomy. There is general weakness, of equal degree, in the whole musculature, to the extent that the patient is incapable of keeping himself upright or pressing firmly with his hand. Both oculomotorii are almost wholly paralysed in all their branches, only the pupils and accommodation are normal. The vision is good. All the special senses and all qualities of sensitivity of the skin are normal. Patient is so sleepy that he falls asleep while being examined, while eating, etc., and can only be kept awake by vigorous shaking. Up to *18 December* the weakness increases further and becomes more pronounced on the right side. The sleepiness continues. *5 January*. Ophthalmoscopically no definite finding, p. perhaps somewhat reddened, more so in the left eye. *8 January*. Total **[242]** hemiplegia on the left side with somewhat reduced sensitivity, continues until *18 January*, on that day it seems to have disappeared; at the same time great agitation has taken the place of apathy, the patient complains of pain in his right leg, his facial expression is somewhat more lively. In the evening slightly raised temperature, for the first time some tracheal râle. The next day is very good, on *20 January* extreme sleepiness again. Varying behaviour until *7 February*, when it is noted that the sensitivity on the right side is somewhat less acute but otherwise normal, that the special senses are functioning and the intelligence is normal once the patient has been woken up. The left pupil is dilated. *8 February*. Ophthalmologically no change. *10 February*. Increasing weakness and weight loss, bladder incontinence begins. Incipient pressure sores are noted on *15 February*, on *17 February* death occurs. In the last month the temperature varied between 37.4 and 38.5, rising when the patient awoke. The urine taken from the bladder after death contained neither protein nor sugar, but upon addition of alkaline copper solution a black precipitation formed, the nature of which could not be determined.

Autopsy. The central cavity grey [matter] of the 3rd and 4th ventricles thickly permeated with capillary apoplexies. The lesion begins at the front, at the anterior commissure, coats the interior walls of both thalami, encompasses the taeniae of the thalamus, the pineal gland, infundibulum and tuber cinereum, as well as the enormously enlarged middle commissure, and also the area around the Sylvian aqueduct and the grey floor of the 4th ventricle as far as the tip of the calamus scriptorius. Apart from the punctate haemorrhages the illustration shows a general yellowish-grey discoloration and a very noticeable injection of the fine vessels. A frontal section of the area of the crus of the cerebrum, just in front of the pons, shows, according to the text, that the lower level -- that of the pes -- has remained healthy, while according to the illustration the left side is for the most part also affected; the lesion, considered by the author to be inflammatory, encompasses the whole transverse section of the tegmentum including the left brachium conjunctivum. Both thalami are affected all over but in an irregular fashion, in such a way that there are parts that are normal in between the affected parts; on the left the lesion seems more significant and also extends to the wall of the lateral ventricle. Microscopic sections of the inner surface of the right thalamus, made using the fresh preparation, confirm the assumption of an inflammatory process. The vessels were carefully examined and showed no lesion.

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FIRST DESCRIPTION OF KORSAKOFF'S PSYCHOSIS (1887)

In 1887, six years after the description by Wernicke, Korsakoff gave the first comprehensive description of the unique amnesic syndrome which is designated by his name – Korsakoff's psychosis (Korsakoff, 1887). The syndrome was elaborated in a series of articles between 1887 and 1891 [Korsakoff (1887); Korsakoff (1889a); Korsakoff (1889b); Korsakoff (1890a); Korsakoff (1890b); Korsakoff (1891)]. His third article, published in 1889, was translated into English by Victor and Yakovlev (1955). These authors felt that this was important because "many of his original observations and ideas have been misquoted, distorted or even completely lost in the secondhand treatment of the subject by subsequent writers" (Victor & Yakovlev, 1955).

DISCUSSION

Little has changed since Wernicke described his disease in 1881 and the diagnosis today is still based on careful clinical observation. We review the literature in a sister paper, also published in this issue (Thomson et al, 2007b). However, an important advance in the diagnosis of WE was made by Caine et al, 1997 when they developed operational criteria to differentiate between Wernicke's Encephalopathy alone or in combination with Korsakoff's Psychosis or hepatic encephalopathy. The criteria for WE require two of the following signs: (1)

dietary deficiency (2) oculomotor abnormalities (3) cerebellar dysfunction and (4) either altered mental state or mild memory impairment. This retrospective study, in which the diagnoses were confirmed at autopsy, showed that using the operational criteria, ante-mortem identification of WE can be achieved with a high degree of specificity although the accuracy of diagnosis was reduced when hepatic encephalopathy was present.

The responsibility of the clinician is to identify patients at risk of WE as early as possible, and to institute effective prophylactic therapy (Thomson and Marshall, 2006a, 2006b). Doctors have come to rely too much on the "classic triad" of signs for diagnosing WE, which only occur in 10% of patients on clinical presentation. However it has been known for many years that other symptoms and signs indicate that the patient is at risk. Despite advances in technology, clinicians still rely on the symptoms and signs described by Wernicke 120 years ago. This is a tribute to his powers of observation and description, and should serve to remind clinicians of the 21st century to preserve their visual acuity, because Wernicke's Encephalopathy is still essentially a clinical diagnosis.

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