WSHoMSoc lectures Dec 8th

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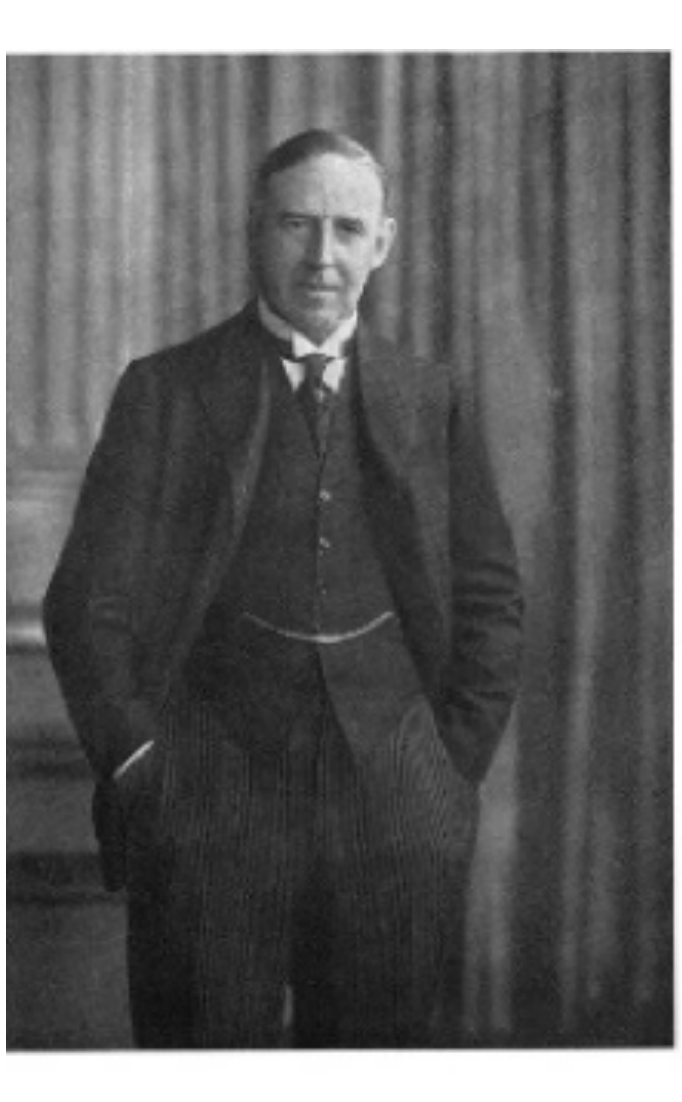

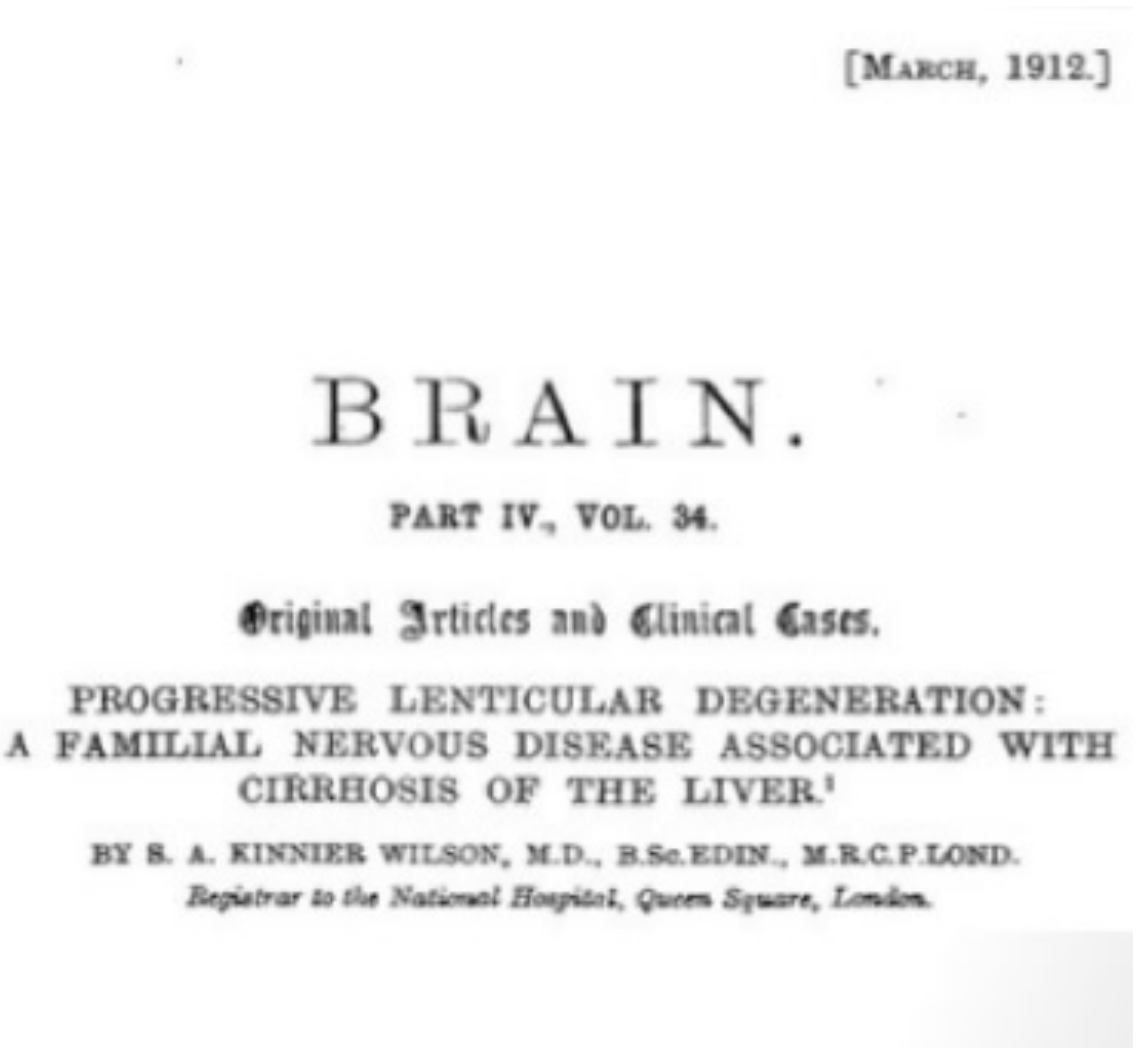

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BRAIN.

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Original Articles and Clinical Cases.

PROGRESSIVE LENTICULAR DEGENERATION: A FAMILIAL NERVOUS DISEASE ASSOCIATED WITH CIRRHOSIS OF THE LIVER.[1]

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Well, another term of fantastic lectures is over. I will miss them. The format is so good: the fortnightly event, the pair of lectures, the local lunch, the elevated conversation… What could be better?

We ended on a very strong note and I for one am agog with anticipation, awaiting the program of 2013 speakers. Let's see if I can do justice to both speakers in this, my humble blog version of todays event. Apologies for any mistakes or unintended omissions.

First off I learned about a luminary of the neurology world of whom I had never heard (at least, I had, but without realising it) – Samual Alexander Kinnier-Wilson.

Our speaker, Dr Edward Reynolds MD FRCP FRCPsych, is a renowned and much published neurologist with a special interest in psychology. His subject was S A Kinnier Wilson of the eponymous Wilson's Disease, who was one of the greatest neurologists of his era, the early part of the twentieth century. The reason for Dr Reynolds coming to Chichester to give this talk was, I believe, the recent unearthing of a film that he intended to show us, but which could not be induced to play on the CMEC computer system due to 'incompatibility', but we were not disappointed as the film was well described and commented upon and I am sure we all felt familiar with it by the end of the talk.

Kinnier Wilson worked at the 'National Hospital for the Paralyzed and Epileptic', a great name for what was surely a great institution. These days it is known as the National Hospital for Neurology & Neurosurgery in Queen Square which is a more fitting name, less like a side-show. (It tends to be called just Queen Square to medics, for brevity.) Wilson was the first person to carry the title 'Consultant Neurologist' and had a great influence on the psychiatric aspects of neurology through his work in asylums.

Wilson's Disease is a rare and pretty obscure disorder and I have not seen a case yet (though I had to ask the neurologist for a ceruloplasmin test recently as I diagnosed an early onset Parkinson's patient who could have had Wilson's Disease, which can present with similar symptoms) and one can see why it interested Kinnier Wilson. Patients have both neurological and psychological symptoms as copper accumulates in their brain and liver. Symptoms such as confusion, seizures, migraine, crazy impulsivity, apathy, dementia and psychosis may be the result of the aberrant metabolism which is due to the absence of the enzyme that breaks down copper. (Please Dr Reynolds, send me a comment and let me know if I have got this *roughly* right, correcting me if not).

But, interestingly, Kinnier Wilson appears to have had a connection with the great Charlie Chaplin! Who would have thought it? The theory was posited that they may have met when Chaplin's mother was in a lunatic asylum, and it is through this connection with Chaplin that it is thought that the film (which in our case we have not got) of neurological diseases featuring Kinnier Wilson and patient examples of neurological defects of many types, came to be made, as Charlie Chaplin would have had the cinematic skills and kit to make such a film. We were shown a photograph of Charlie Chaplin standing with Kinnier Wilson's wife Muriel (nee Bruce) at his large Californian home, and we suspect that Wilson himself is taking the picture, but we dont know who owned the camera, Wilson or Chaplin. It was suggested that the famous walk adopted by Chaplin in his portrayal of the penniless unfortunate that became his alter ego, was developed through his observation of patients in Kinnier Wilson's neurology wards.

Incidentally, I have an interest in epilepsy, not as a neurologist but as a clinician and clinical nutritionist. Many of my readers will be aware of the great efficacy of a ketogenic diet in most forms of epilepsy. The ketogenic diet is very low in carbohydrates, very high in fats and with normal protein intake. This dietary approach was known to be highly effective in treating epilepsy prior to the advent of anti-epileptic drugs such as sodium valproate (epilim) and it has been the focus of renewed interest in the research community of late, research that I am reading and applying, along with a swathe of other clinicians determined to improve the lives of those who experience seizures. In fact the use of such a diet has now been shown to have significant effects in virtually *all* common conditions of the nervous system including brain injury, stroke, multiple sclerosis, schizophrenia and Alzheimer's and Parkinsons, to name but a few. Oxford University is currently recruiting for a trial of the ketogenic diet in Parkinson's disease following a small but very successful study by Theodore VanItallie in 2005. Since it is also extremely effective in disorders of metabolism such as diabetes (type 1 and 2), polycystic ovary syndrome, acanthosis nigricans and obesity (in fact a recent Cochrane Collaboration meta analysis concluded that it is *the most* effective diet for weight management in the long term while simultaneously *improving* cardio-vascular risk markers). The ketogenic diet also has the abilty to reduce the activity and aggression of cancer cells by restricting glucose, as per the Warburg Effect. (This was named after Otto Warburg who was awarded the Nobel Prize for physiology in 1931 for his work on the shift seen in malignant cells to anaerobic respiration of glucose, rather than the healthy aerobic oxidation of fatty acids in the Krebs Cycle in non cancerous cells).

I was extremely touched by the letter we were shown, written so elegantly by Charles Sherringham to Kinnier Wilson telling him that he and Edgar Adrian had proposed Wilson for the honour of Fellow of the Royal Society. The letter was filled with gentility, savoir faire and Edwardian courtesy. The handwriting alone was a delight. I loved the way Sherringham entirely down-played the fact that he and Edgar Adrian had just got home from receiving their Nobel Prize for Medicine by writing something like: 'Adrian seems no worse for wear after his trip to Stockholm'. How very tangentially put! That was the era when blowing ones own trumpet was simply vulgar. These men were anything *but* vulgar.

The second of this last pair of lectures was by our very own Winston Leigh BA MB ChB MRCGP. I say 'our very own' because this doughty gentleman has now given no less than eight lectures to members of the West Sussex History of Medicine Society. He certainly gets the prize for persistent curiosity. (I hope I get one of those when I grow up too).

Winston Leigh must be an artist, or a musician, or both. I have only been at two of his lectures so far (I have only been a member of the society for a couple of seasons) and both have been about artists and had music playing as we went back into the lecture hall after our tea. (Thank you to the elves that make the tea and coffee appear as if by magic, by the way!) This time the strains of a Robert Schumann piece met me as I found my seat and I am afraid to say I mis-guessed the composer.

The subject of the lecture was Artists's Eyes: Oscar Claude Monet and Vincent van Gogh. We were lead through the various theories and their propbable accuracy about the distinct artistic styles that these great painters are famed for and whether their eyes, their brains, or simply their painterly idiosyncrasies were the source of their genius.

It seems that there is a new sport which medical minds spend considerable time and effort on. It involves trying to diagnose the illnesses of historical figures in today's terms. Eyewitness accounts, records of one sort or another as well as things like CT scans, bone analysis where possible or tissue sampling can lead to a more accurate appraisal of their diseases, so it is a rich and fascinating seam indeed, there being a great many historical figures to investigate! (For example: Fredrick Chopin may not have died from TB, as has been generally assumed, but actually have had cystic fibrosis? That was the subject of a previous lecture by Dr Leigh last year and very convincing it was too).

Seeing a picture painted in 1867, then one in 1890 and another in 1922 was a wonderful way to witness the marked change in Monet's style, as it did indeed become more and more 'impressionistic'. But was there a cause, per se? Indeed there may well have been: we learned that Monet certainly developed cataracts, in both eyes. By the time he was 53 and living at Giverny, enjoying his fabled garden, his colour differentiation was definitely impaired, which must have been a cause of great anxiety to him, though great pleasure to those of us who love his water lily works. He very sensibly rejected the idea of surgery for many years, but eventually did risk it in 1923, when it had to be done in two stages as he apparently became aggressive and uncooperative with the surgeon. Poor man! He must have been terrified, and not without reason. I know it is a very common operation these days, but most people still balk at the idea of letting anyone near their eyes with a sharp implement. I learned the word 'aphakic', (thank you Dr Leigh) as he had to use aphakic glasses once his cloudy lens was removed.

Here are two examples of his work, two views of the same bridge and pond painted 22 years apart:

On A Serene Spot By A Pond 1897

The Footbridge Over The Water Lily Pond 1919

Vincent Van Gogh is quite a different case. It seems that his physical and mental health were the main problems, mostly brought on by poverty and subsequent poor living conditions, and he was known to have had epilepsy which *still* leaves many at a social and financial disadvantage. However he smoked a pipe and drank heavily too. It seems too that he was acquainted with the Green Fairy (absinthe) which was a common drink amongst the bohemians of the era. As a Medical Herbalist I have prescribed tincture of wormwood (*Artemisia absinthium*) on many occasions and can honestly say I have never seen anything but excellent effects when used for its vermifuge and anthelmintic properties with no wacky side-effects in the least, so my belief is that the virtually hallucinogenic properties ascribed to the drink absinthe were exaggerated and played upon by users and detractors alike. He also had gonorrhea and suspected syphilis too, possibly from the alcoholic prostitute he shacked up with for a couple of years. So he was not a well man.

However, our speaker was interested in the artistic style, use of colour, especially the famous yellows, and the halos he painted around lights and stars and whether all this added up to a quirk of the visual cortex or other apparatus, or some other discernible pathology. However, in the final analysis it did not. His eye was fine, and no sight issues have been revealed. He seems to have had a particular affinity for a mustardy yellow colour, using various shades intensively in a great number of paintings. One amazing piece called Still Life With Grapes, Pears and Lemons is painted virtually entirely in hues of yellow. He lived in a house called The Yellow House, whether by design or co-incidence, but none of his use of colour can reasonably be considered pathological anymore, but rather part of his artistic genius.

Apart from the extremely interesting thoughts about Van Gogh's health the culmination of Dr Leigh's talk was, for me, the wonderful news that Van Gogh probably did not die due to an unsuccessful suicide attempt using a gun as we have been lead to believe. We were reminded that the original story goes like this: on July 27th 1890 Vincent was in a wheat field in Auvers-sur-Oise with his easel and paints, working away, when his poor mental health got the better of him and he shot himself. Regaining consciousness and with a chest wound he struggled back to the hotel in which he lived. Two doctors were called, and word was sent to his brother Theo, but the doctors did not have the skill to remove the bullet which miraculously had missed his vital organ but was lodged towards his spine. Vincent stated that he had pulled the trigger himself and that no one else was involved. However, it was known that he had taken to hanging out with two young chaps, brothers, perhaps in their late teens or early twenties, and together they would drink and get sozzled and have good time (presumably). These brothers had a revolver and it is now believed that on this fateful day the three of them were in the field together, considerably the worse for wear due to alcohol, and one of the brothers accidentally discharged the gun inadvertently wounding Vincent! They probably scarpered, with the gun (which was never found) assuming him dead and terrified of the consequences, but he was only unconscious. Once back at the hotel he clearly stated that it was a stupid attack on himself by himself and that no one else should be considered, and he smoked his pipe and waited for Theo to come and sort him out (again). Overnight however, the wound became septic, which is to be expected in an emaciated, thoroughly malnourished body, so the next day he died! But the wonderful news is that some thirty-odd years later a middle aged man identified himself as one of the two brothers who had been with him on that drunken day, and that he had indeed been the one to have actually pulled the trigger of the dratted gun, by accident! What a hero it makes Vincent! He made sure that the young chap was not hounded as a murderer, as it had all been a stupid, idiotic, clumsy accident, and nothing would be gained by blaming the lad. He certainly does not sound insane now does he? More like an entirely decent if intensely foolish genius artist, sick in body like so many other impoverished folk, but which the world would have been so much the poorer without.

So all those possible diagnoses that Winston Leigh offered up, but discarded, such as *bipolar disorder* (apparently his sister 'had' it), *Meniere's disease* (which could explain the ear incident, by the way it was definitely his LEFT ear – don't be fooled by the self portrait wherein it appears to be his right ear – he was looking in a mirror while painting it, duh..), *closed-angle glaucoma* (the halos round the stars and candles in his works, but in a man not yet 40 this would be unusual), or *neurosyphilis* (again, he would have been rather young for it to have manifested neurologically), nor *Digitalis toxicity* (though this could have been the cause of the xanthopsia – seeing excessive yellowness – as Dr Gachet of the eponymous painting by Van Gogh did prescribe foxglove frequently and is painted holding a sprig of it, but there is no evidence that Vincent had a heart arrhythmia nor was the recipient of that particular herbal drug). So we are left with simply the wonderful, rich, vibrant, startling and vivid painterly intention, rather than any ill health 'causing' Van Goghs colour choices.

Haystacks in Provence June 1888 Arles

Briefly, I will now add *my* opinion on some of Van Gogh's medical history, for what it is worth:

Epilepsy is cured or markedly improved in many cases, children and adults, including some of my own patients, by the adoption of a ketogenic diet. A ketogenic diet, as mentioned earlier in this post, is one in which dietary carbohydrates (which all turn into glucose in the blood, which in turn causes a spike in blood insulin) are kept to a daily *maximum* of 20 – 30 grams, i.e a tiny amount, and those lost calories are *replaced* by the ingestion of more fats. This caused the liver to produce ketones which stabilize neuronal firing, and thus seizure frequency. (Of course there are highly complex biochemical effects too, but in short that is what happens).

As we see from the life of this great artist he was poor, dirt poor, much of the time, relying on his brother for funds, and therefore could only afford the cheapest foods, which are always the bulk carbohydrates such as potatoes, wheat or bread, and possibly root vegetable soups. The more nourishing foods such as eggs, meats and cheeses are always the more costly foods and are made up of fats and proteins. He only sold one painting in his lifetime, and lived amongst peasants, i.e. the poor.

One of his early paintings, *The Potato Eaters* is particularly poignant to me, as clearly there is very little nourishment, in terms of protein or fat, in the humble spud. It is almost entirely a source of starch, which rapidly turns to glucose in the blood. Living on starch will ensure a short life, and poor quality offspring, and the possibility of neuronal disturbance and low IQ as a result, just as we see in his depiction of those effects. He also painted wheat fields frequently, and the hard, back breaking work of sowing and reaping the grain that peasants had to do. I think this is best called 'scratching a living'. Other mental conditions which in some cases appear to be significantly improved on a ketogenic diet include: Obsessive Compulsive Disorder (OCD), Bipolar Disorder, Depression, Schizophrenia, Migraines and Autism Spectrum Disorders, and those mentioned above. Here are a couple of links to two recent papers (sourced from PubMed) to whet your appetite regarding the progress being made in this area of nutrition and metabolic health research:

1) Kraft BD and Westman EC. **Schizophrenia, Gluten, and Low-Carobhydrate Ketogenic Diets: a case report and review of the literature**. *Nutrition & Metabolism 2009, 6:10*

2) Phelps JR, Seimers SV, El-Mallakh RS. **The ketogenic Diet For Type II Bipolar Disorder**. *Neurocase Oct 3rd 2012*

So, if I had been asked for health advice by Mr Van Gogh, made possible due to a time slippage incident, my first recommendation would be that he eat food of a higher nutritional content i.e. rich meaty stews, fish of all sorts, omelettes, cheeses and hams, and nuts or olives in between if needed. I would encourage him to include plenty of fat in his diet, fatty meat, high fat cheeses and oily fish, and that fat in the form of butter, or olive oil be added to everything he ate. And I would have got him to have full fat plain yogurt daily too. And I would certainly get him off potatoes and bread and that sort of high carbohydrate, padding food, worthless peasant fare that is low in nutrients, vitamins, minerals and general food value. I would have tried to get him to stop smoking and to markedly reduce his alcohol intake, but only once his food had been corrected, as that would make the biggest difference to his health. And I wouldnt want him to lose his muse now, would I…?