OS17 **GCA** Tapering is an inexact science

Looking back nearly three years it was like a sick version of the joke about how many actors / Jewish mothers / psychiatrists etc it takes to change a light bulb. In this case, it was: How many doctors does it take to diagnose a case of Giant Cell Arteritis?

The answer was none. In my case, it was not a medical expert who came up with the right answer but a friend.
It all started with a headache that got increasingly worse. Over a two month period in which sleep was reduced by pain to two hours a night, there were consultations with two GPs, a specialist, a brain scan and day surgery, all of which did nothing to help. Finally an old friend came up with the correct diagnosis: “I knew someone with symptoms like yours”, she said. “It sounds to me like Giant Cell Arteritis”.

I had never heard of it – and neither, to terrible cost, have most people in Britain, including it seems from my experience, an awful lot of doctors. But she was right. And she probably saved my sight, maybe even my life.
My experience, I gradually learned, extreme as it may seem was not exceptional.

Because most people know nothing of the condition and its seriousness when it strikes, and because many doctors simply don’t recognise the symptoms or, if they do, do not treat them properly, about 3,000 people become blind needlessly very year. It is the most common cause for unnecessary acute vision loss in this country.
I later talked to to one of the world’s leading experts on the condition, consultant rheumatologist Professor Bhaskar Dasgupta, of Southend University Hospital, who told me that victims often have clear symptoms before blindness strikes and could be spared if only their doctors acted promptly and accurately. “The problem”, he says, “is that both public and professional awareness are very low.” As, in effect, a stroke in the eye, it should be treated as a medical emergency but all too often it is not.

GCA hits about 13,500 people every year, two thirds of them women, and almost all of them over 50. And there probably lies the rub: the condition has suffered low priority because of ageism: “If elderly people do go blind, it’s seen as more tolerable”, says Professor Dasgupta. “If they were all younger, it would get greater priority”. There is also the fact, say some, that many sufferers belong to a generation brought up “not to cause a fuss” and too often regard some symptoms as those of getting older. As it is, say experts, research into and treatment of GCA is years behind that of other conditions. “In knowledge and in practice, we lag years behind other medical conditions,” says Professor Dasgupta.

Over the month I found out a lot about GCA. The name – it is also called temporal arteritis because it commonly, though not exclusively, affects arteries of the skull - acknowledges the abnormal large cells that develop in the wall of the inflamed arteries. It is an autoimmune condition: the immune system instead of carrying out its usual protective role attacks healthy arteries. (It is called arteritis because it attacks arteries, not veins.) No one knows what causes it – though unknown infection might be involved and although it is not directly inherited, genetic factors may play a part. Poorly researched, little is known about it - including whether it is a disease of modern times or not (there are unproven claims it might be been seen in Egypt as early as 1350BC). There is no cure.

The good news, as sufferers know, is that it can be controlled – and in most cases sight can be saved if treatment begins quickly enough. The bad news is that the treatment can sometimes seem as bad as the condition itself.

Although GCA can seemingly strike hard from the blue, there is often a build-up of symptoms, too often regarded by the sufferer as inevitable bi-products of aging and by doctors as signs of other more familiar conditions. In my case it began with a mild flu-like

feeling which I attributed to overwork. Then pains began in my neck and travelled up to my head. As it worsened, sleep fell to one to two hours a night. My GP prescribed painkillers, first ibuprofen, then more potent ones. The pain settled over my right temple, a symptom that I learned later should have been a giveaway. A second GP thought it was a complaint called trigeminal neuralgia, a nerve disorder that causes pain so great it is sometimes called the “suicide disease”. Ordinary painkillers are useless against it, so a third, more specifically targeted drug was prescribed. I was also referred to a pain consultant who organised a brain scan (in case of a tumour).

The consultant prescribed yet a fourth drug, and decided I needed day surgery for nerves to be anaesthetised for a short period. The rationale, as I understood it, was that he believed the nerves were having some kind of frenzy; being put to sleep for a while would allow them to calm down. Bliss: for 24 hours the pain vanished – and then returned with a vengeance. By now, I was permanently exhausted (I didn’t know how much was due to lack of sleep, how much due to whatever my complaint), had lost four kilos, and had lost all desire to eat. Then came the bouts of blurred and double vision. And, thankfully, the meeting with the old friend. I mentioned my friend’s thoughts to the consultant. “Ah yes,” he said, “I had been wondering about that.”

Twenty-four hours later I was with a rheumatologist who, at 7pm, sent me for hospital blood tests. Three hours later he telephoned me at home telling me to be waiting on my GP’s doorstep the following morning with his mobile phone number: the hospital had called him because the test results were “grossly abnormal” and in line with GCA. I needed immediate treatment.

By 10am the following morning I was swallowing my first mega dose of the corticosteroid prednisolone, the beginning of a long regime which still continues. The potential consequences of not acting immediately if GCA is suspected are so great – notably blindness and more rarely stroke – that treatment should begin even before the doctor can be certain.
This has been a major cultural problem for many GPs whose usual practice is a cautionary one of proceeding step by step, not leaping in with huge doses of a drug notorious for its side effects.

The blood tests I received - erythrocyte sedimentation rate (ESR) and C-reactive protein - are ones that detect inflammation and if high are strong indicators of GCA. Confirmation comes later – usually, as in my case, with a small operation to remove a small section of the Temporal artery, and to examine it under a microscope where the giant cells are revealed eating away at the artery and blocking it as well.

From that moment the high dose continues for a month (relief comes within days), and is then tapered over many months, even years. In some cases a small dose of prednisolone needs to be taken for ever. Tapering is an inexact science, developed by experience, and meant to balance two risks – that of the GCA and that of the drug keeping it under control. Some rheumatologists taper more slowly than others, believing the damage being caused by the steroid is balanced by the patient suffering fewer relapses which mean having to increase the steroid. It usually takes 1-3 years to come off steroids; the average time in one study was 22 months. Because the body’s adrenal glands cease producing their own steroid, it is dangerous to stop them suddenly and users have to carry a steroid warning card in case they are involved in accidents.
The most important fact about it is that it works. In the case of GCA, the effect within two to three days is near miraculous: headache gone, energy returning.

Both GCA and its treatment also need other drugs to be given in concert. I found myself taking a handful of other pills: low dose aspirin adds to the protection of vision; calcium and vitamin D help product the bones, a bisphosphonate also reduces the risk of

osteoporosis, a tablet called a proton pump inhibitor helped protect the stomach.
As one sufferer said, “Having GCA is no fun; nor is taking the steroids, but it’s better than the alternative.”