

WHO WERE CHARCOT, MARIE AND TOOTH?

By Paul Cooke

Jean Martin Charcot

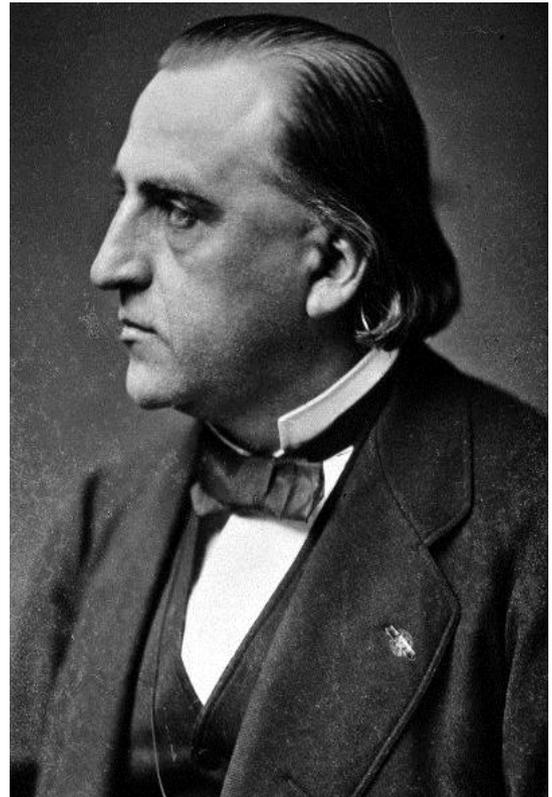
1825-1893

A Study of success against adversity

The story of John Martin Charcot is surprisingly the story of a man who rose from poverty without advantage, living through appalling conditions of war and revolution.

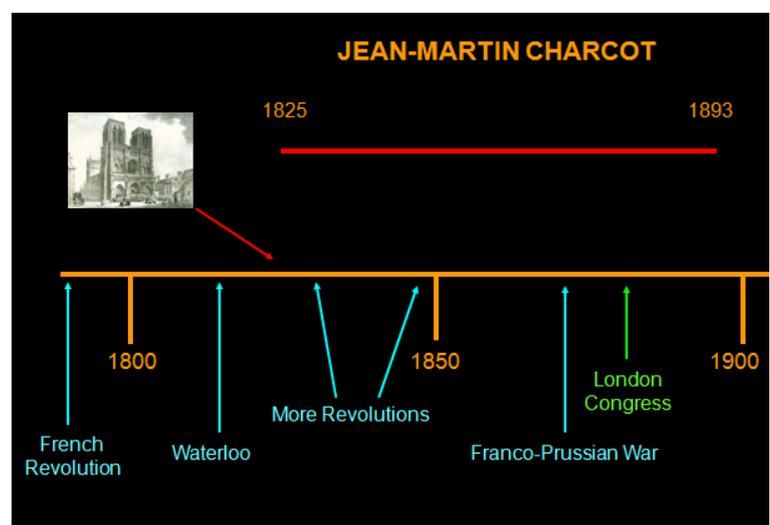
A story which I hope will entertain and inspire you.

I will describe in turn his early life, his medical career, his achievements, and his teaching and famous pupils.



In order to put things into context, here is a time line.

You will see that Charcot was born after the first and main revolution in France, between the French defeat at Waterloo in 1815 and a further revolution in France of 1830. Before studying Jean Martin Charcot, I was blissfully unaware that there were so many French revolutions – although fans of “Les Miserables” will be more aware of this.



He lived through a turbulent time in France - two revolutions and a war -but was able to attend the International Medical Congress in London in 1881.

He died just before the end of the nineteenth century.

He was born in Paris on November 29th 1825 to a family of very modest means.

His grandfather and father had both worked as saddlers and carriage builders, and he came firmly from the working classes of the times.



His father had high ambitions for his children, but given his limited means, he realised that he could only afford to sponsor one of his four children to pursue a learned profession.

The family organised an assessment of intelligence and achievement for each child, and Jean-Martin was assessed to have greatest potential.

So his education continued whilst his three brothers left education and went on to the army, to sea and

to coach building respectively.

As a child and youth he was variously described as taciturn and introspective, and it was recorded that he liked to read and to draw, and showed special talent at this latter activity.



We have few examples of his early drawings, with the exception of an early sketch of a professor or teacher, but his skill at drawing was noted in school records

He attended the “Lycée” Bonaparte – the main secondary school in Paris - before progressing to university study in Paris.

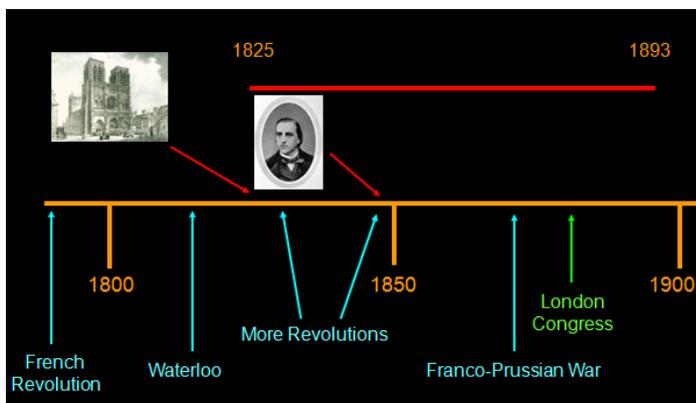
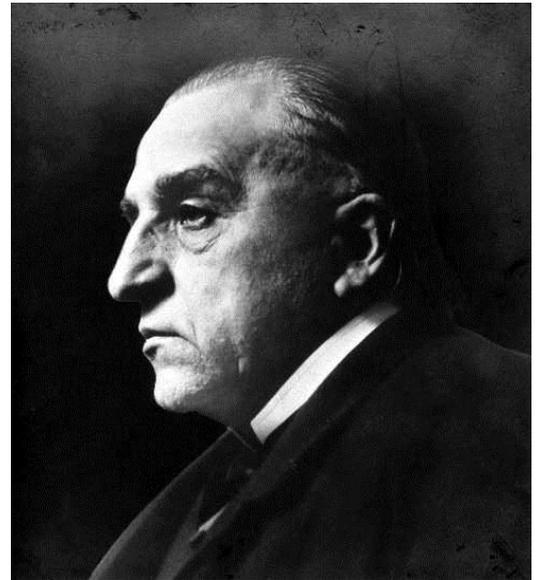
There appears to have been agreement within the family to help him achieve, and the brothers showed extraordinary fraternal support by helping him study whilst they

attended to his other duties and his needs – including keeping his study room warm in winter by carrying heated cannon balls in, which, when rested in buckets of sand, acted as radiators. I wonder how many of us would have done that for younger brothers?



Thus supported, he went on to complete his schooling – at the end of which he had to decide whether to follow art or medicine as a career. Fortunately he chose medicine.

Charcot embarked upon his medical career from 1848.



This year, 1848 saw yet another revolution in France.

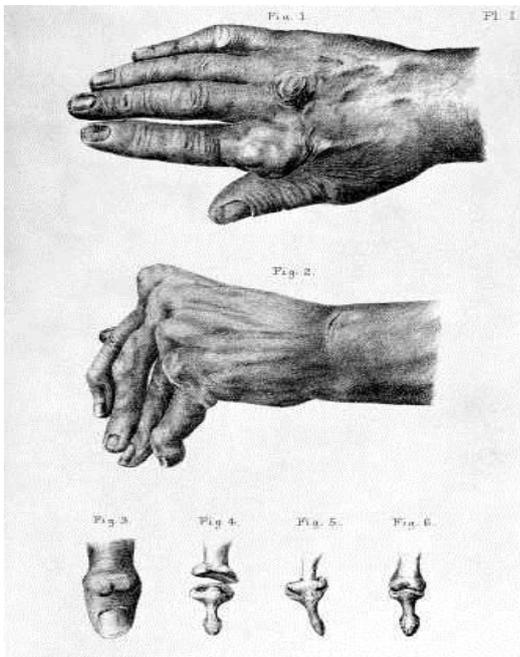
This deposed the monarchy, which had been restored in 1830, and led to the creation of the second Republic.



At the age of 23 he successfully completed competitive examinations for a hospital internship at the Hospitiaux de Paris at the Salpêtrière and instead of starting in general practice, as was usual in England at that time, he went straight into hospital practice.

At the Salpêtrière, he was fortunate to be sponsored by the dean.

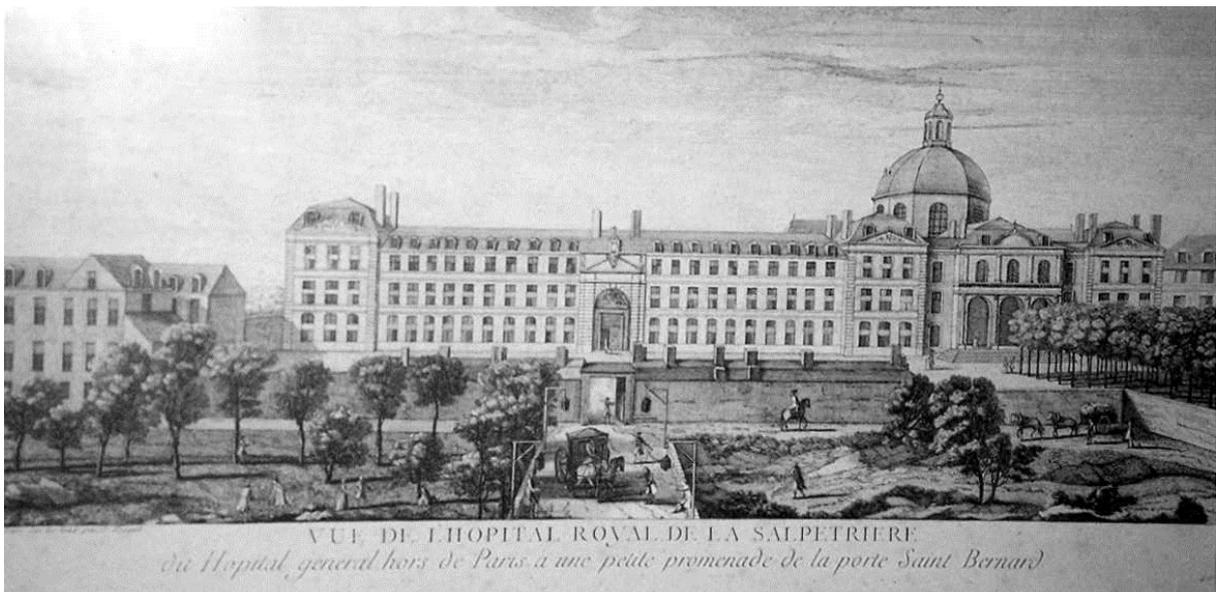
Five years later, in 1853, Charcot presented his thesis on the differentiation of gout from chronic rheumatoid arthritis, which were then often confused as different presentations of the same disease.



At the top you can see Charcot's own drawing from this thesis of a hand with a gouty tophus over the index finger metacarpophalangeal joint;

Below is a hand with typical rheumatoid deformities beautifully depicted;

At the bottom are the pathological changes found on post mortem dissection, gout on the left and rheumatoid on the right all these drawn by Charcot himself.



Hôpital de la Pitié-Salpêtrière

Three years later, Charcot was appointed Chef de Clinique- the clinic boss - at the Salpêtrière .

This hospital was built over 200 years before in 1634 at the instigation of Louis XIV on ground called "Le petit Arsenal", the site of a gun factory - Salpêtre of course means saltpeter, used for manufacturing gunpowder.

The hospital was expanded in 1684 and became a dumping ground for the poor of Paris. It was notorious for its large population of rats.



It is now a general teaching hospital where many celebrities have been treated. Diana, Princess of Wales, died there in 1997.



At the time of the Revolution the Salpêtrière had become the world's largest hospital, with a capacity of 10,000 patients plus 300 prisoners.

It was stormed by the mob in 1792 and many of the female patients were killed in what is illustrated here as “The terrible massacre of women at the Salpêtrière”. This reflects the attitude of the doctors and public to women, and the ill at the time

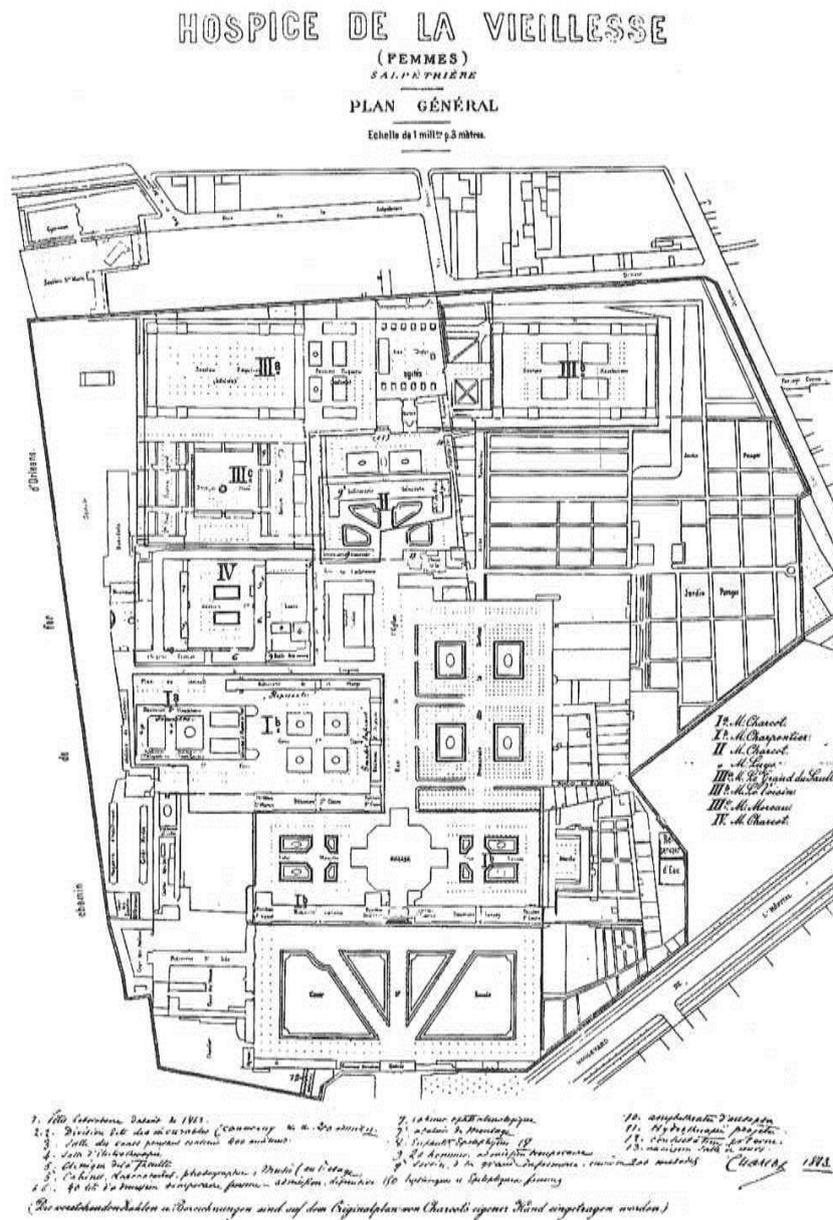
By the early nineteenth century the Salpêtrière was described as “resembling a Hell and a Mecca of female death – but with the incongruous addition that it was a mecca of female death with a splendid cruciform church in the centre”.

When he took over its patient population was approximately 5000, with 1500 insane persons and 2600 disabled and epileptic women, who served as a valuable resource for Charcot’s observational research.



The cure rate then is estimated to have been less than 10% and 254 women died there of causes presumed to be due to insanity.

Remarkably, these were recorded to include masturbation, scrofula (which is tuberculosis of the neck glands) blows and wounds, debauchery and licentiousness, cholera, erotomania, alcoholism, rape, love, joy, bad reading habits, nostalgia and misery. How these were all attributed to insanity or death is difficult to understand now. Maybe that's due to our greater understanding now – or maybe that's just the French for you!



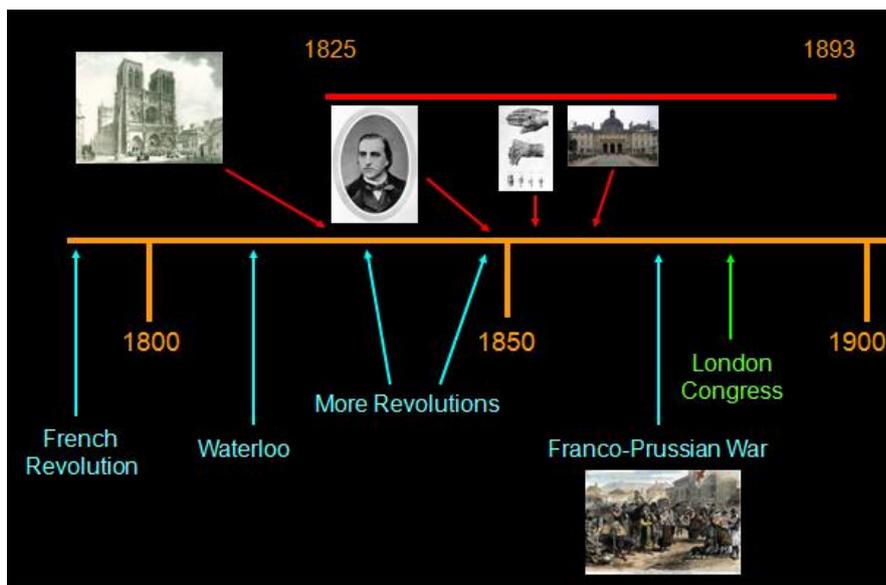
Plan of the Salpêtrière with Annotations by Charcot

Here you can see a contemporary plan with handwritten annotations by Charcot showing his plans for reorganisation.

Charcot led the transformation of the Salpêtrière from a run down poor house into a modern hospital.

He started by creating a laboratory with microscopes in an abandoned kitchen, followed in the next years by the installation of a laboratory for anatomy and physiology, a room for photography and a museum for pathology.

His responsibilities included diagnosis and classification of patients with unidentified diseases, of which there were a large number at the Salpêtrière.



The Franco-Prussian war of 1870—71 and the revolt of Paris (Commune de Paris) which followed interrupted his development of the hospital.



In the chaos of the siege of Paris he spent his time treating epidemics of typhoid and smallpox, which were rife.

As you can imagine, his life was severely disrupted by these catastrophic events. He sent his family to safety in London for the duration. But despite all this he continued rebuilding and gathering clinical data, and was made professor of medicine and neurology in Paris.

So what did he do which made him so famous and led to the award of all these chairs.



Charcot's achievements were so numerous that I can only describe a few.

He was obviously a brilliant physician, but he was also a skilled and unbiased observer who in the environment of the Salpêtrière hospital, was able to observe many mental and physical disease processes.

Here was a case of being the right man in the right place at the right time.

The man was right and hungry to discover. He was the first physician to fight for treatment for female patients, and at the Salpêtrière. this gave him a huge reservoir of patients to study.

The time was right. Because a new way of studying patients was developing, relating clinical observations to outcome and disease



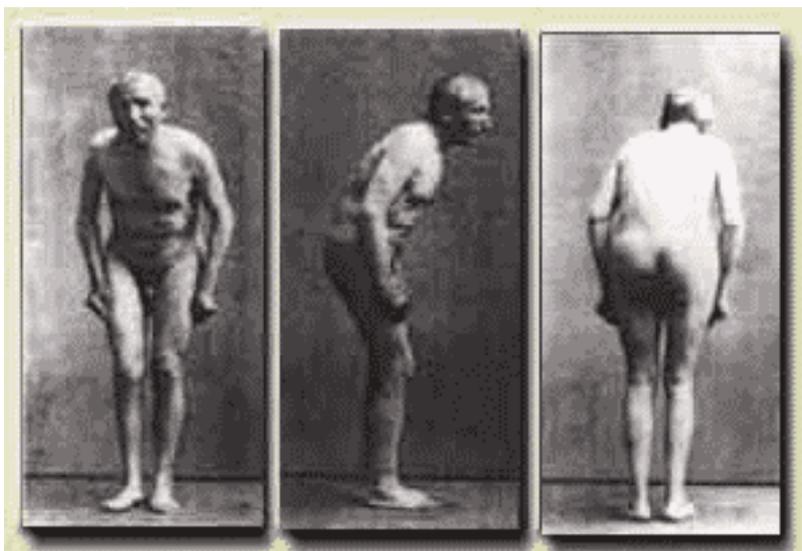
So the time was right.

And of course he was in the in the right place, and he developed this advantage.

He had no shortage of clinical cases of all sorts of disease to study and draw in detail whilst the patients were alive and incarcerated within the Salpêtrière- and when they died – as most did- few had any relatives waiting to claim their bodies and a right to early burial. So there was a ready and probably unique supply of post-mortem specimens from patients who had been studied, observed and drawn in life.

He grasped the opportunity to correlate the clinical observations with the post-mortem findings in a unique manner and told us more about the cause and effect of disease than anyone who

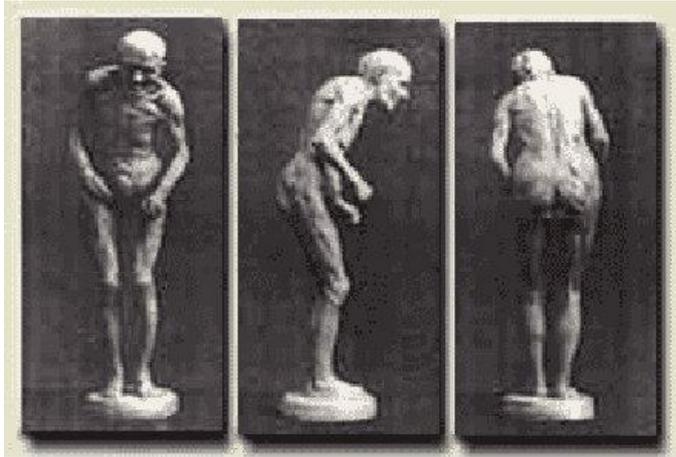
went before.



In life he described their condition in words, he drew illustrations himself -although he later acknowledged the superior skills of Richter who took over this task.

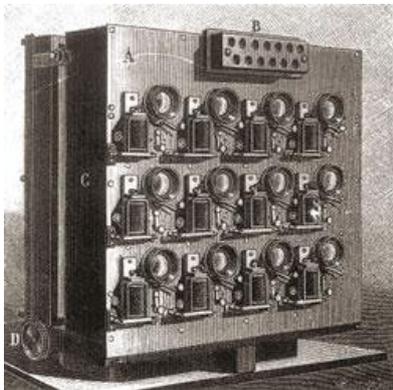
Together they enveloped the developing technology of photography and often made sculptures of patients based on the photographs.

Here we see an example of this discipline of accurate recording, with the posture which leads to the stumbling festinant gait of Parkinson's disease depicted above in photographs – later converted to the statuette below.

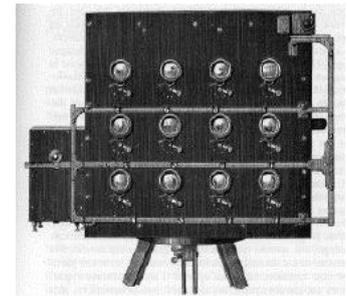


They were not of course content to just use this technology, they developed the new methods of photography as they went – and for this purpose they appointed and encouraged Albert Londe.

Amongst other achievements he developed firstly a nine, then a twelve camera system to show movement.

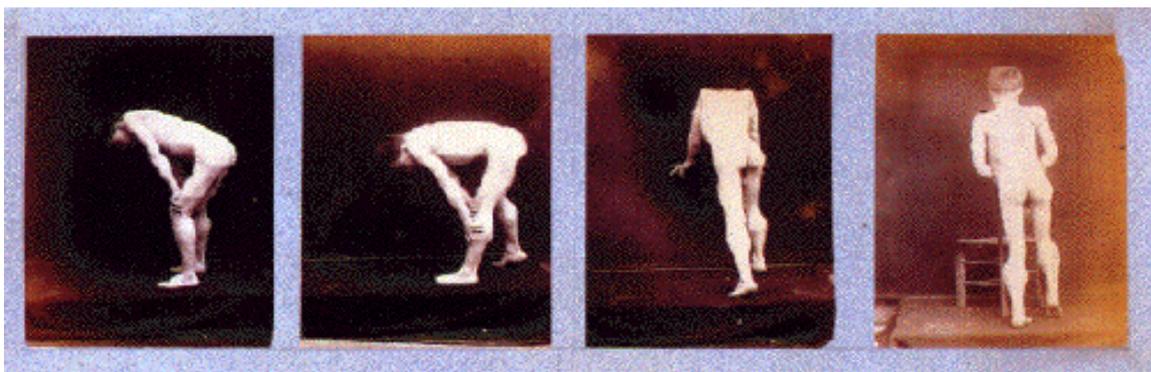


Sequential pictures were taken – their speed controlled by a metronome and we see below a sequence from it showing the classic rise to standing of a child with Duchenne dystrophy.

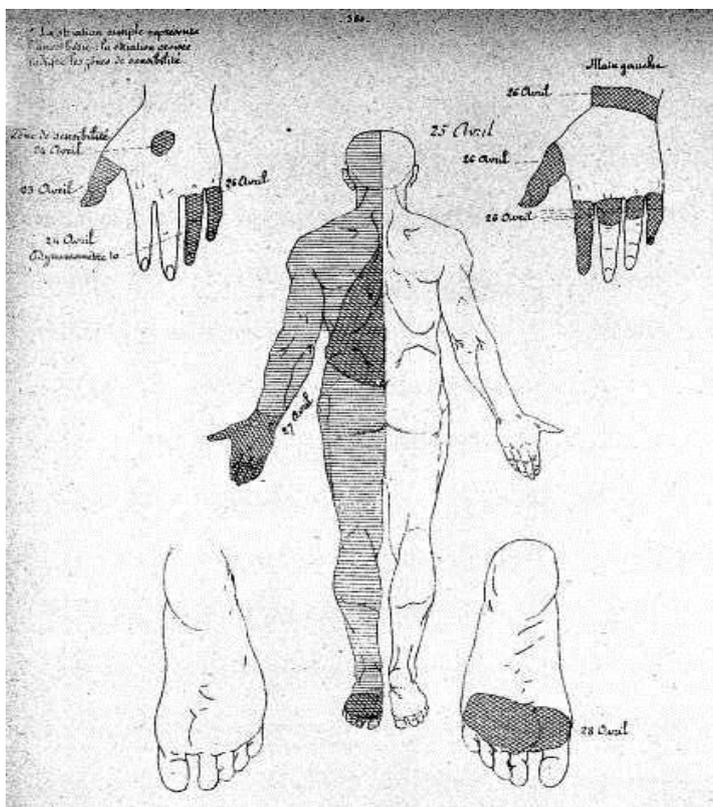


Albert Londe, working in Charcot's department is widely held to be the first

man to photograph movement and as he developed projection methods for these moving pictures is hailed as the forerunner of movie photography and modern cinematography. Who would have thought that it was Charcot had started and encouraged this?



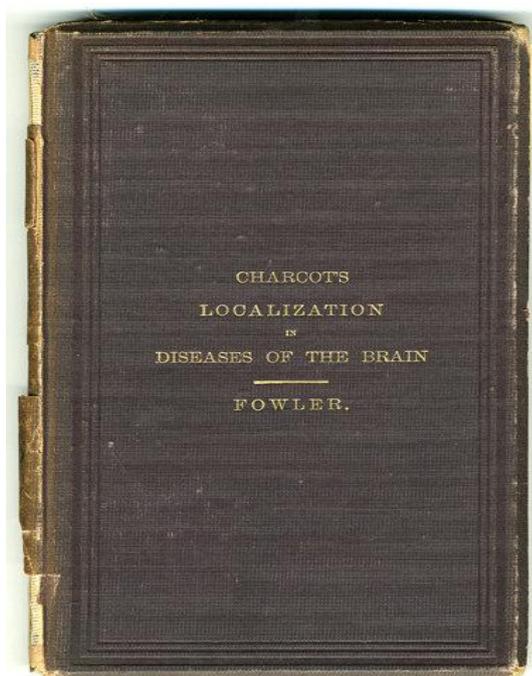
Using techniques of recording by drawing, photography and modelling and correlating these with meticulous clinical records, he also made remarkable discoveries. In three lectures in 1868 on disseminated sclerosis (which we now call multiple sclerosis or MS) he clearly delineated this disease for the first time and described the main neuro-pathological findings.



Here you can see his drawings of the anatomical distribution of the sensory loss.

he was then able to group his patients with tremor into those with intention tremor (whose hands trembled when they tried to do things) and those with resting tremor.

He noted that those with intention tremor had sclerotic plaques in the brain which he found when they died, and this allowed the first differentiation between multiple sclerosis and Parkinson's disease.



So let's look at a few of the subjects he excelled at.

In neurology, arguably his greatest contribution- and that of most relevance today was his anatomical study of the brain.

Initially he studied anatomy – as a proponent of the developing theory that the brain was not just one homologous organ – but comprised different regions with different functions, and he specifically observed bleeding and aneurysms within the brain as a cause of stroke.

His anatomical studies did much to define the areas of the brain and the cause and effects of intracranial haemorrhage and thrombosis, work which has

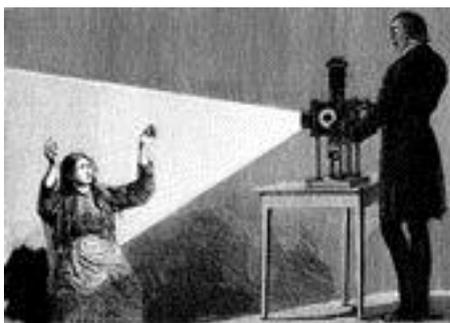


remained as accurate today as it was then.

His contribution to this subject is remembered in the eponymous Charcot artery to the brain, which all medical students learn about and the Charcot Bouchard aneurysm shown on the left.



In mental disease he is most celebrated for his work on hypnosis – leading it from the stage act of mesmerism into a therapeutic modality, although he came close to returning it to a stage act with his public demonstrations and use of lighting to dramatise the effects.



He studied hysteria and is credited by psychiatrists as the first to demonstrate a relationship between psychology and physiology .

In 1887 he was coauthor of a book entitled “Les Demoniques dans l’Art” with Paul Richer.

This contains many monochrome plates to illustrate the subject, including this one of hysterical club foot.

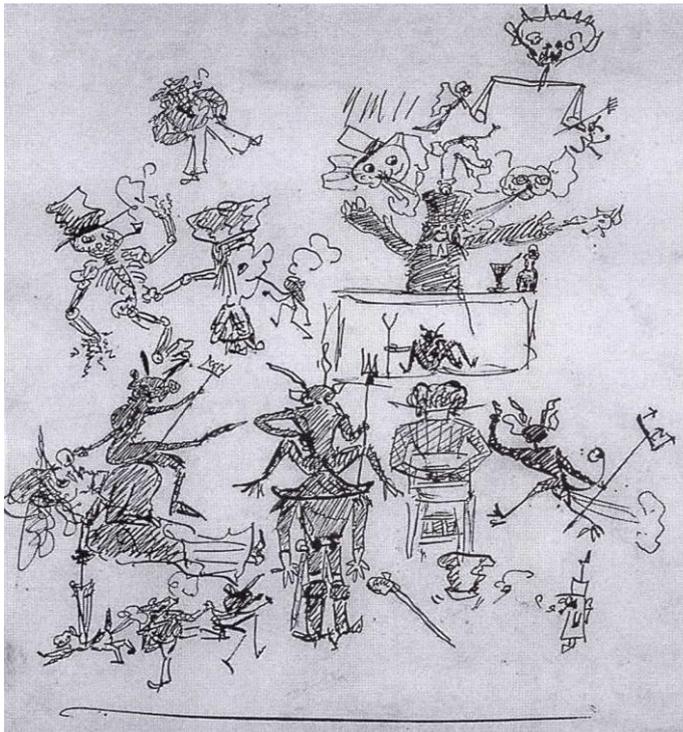




In 1890 he established a Laboratory of Psychology.

In 1893 he wrote a second dissertation on "L'État mental des hystériques"- hysterical disease.

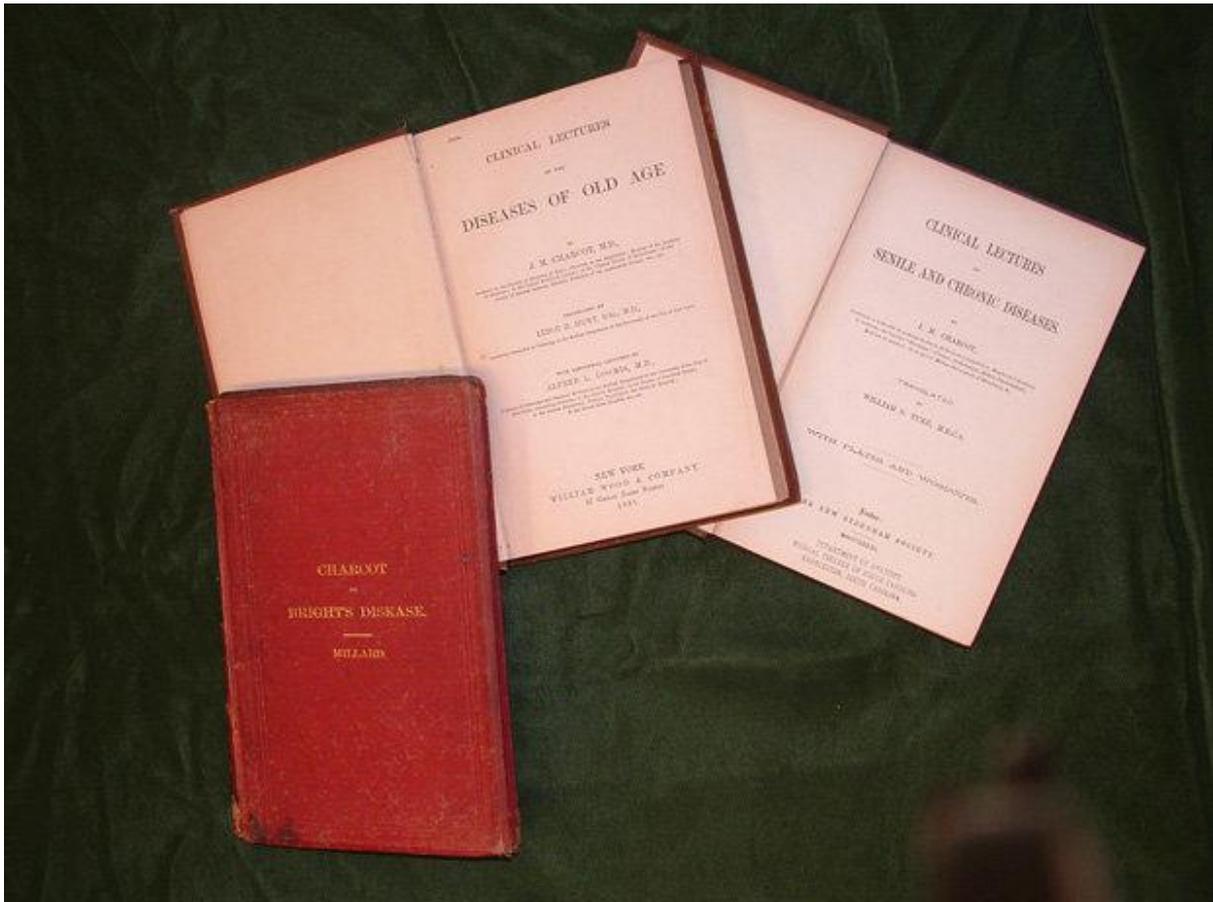
Here you can see an illustration from the book of the insertion of a needle through the arm apparently painlessly in a hypnotised patient.



He tried to understand the workings of the disturbed mind and probably went a bit too far in doing so – taking drugs to induce mental disturbance. In such states he produced disturbing images such as this to illustrate the demons which occupy the diseased mind



He also initiated theories of the sexual origins of mental disease with his pupil Sigmund Freud – who of course went on to develop and refine these. Again how many here would make this connection?



He is celebrated for his remarkable findings in mental and neurological disease but also made remarkable discoveries affecting general medicine, surgery and of course orthopaedics.

In general medicine he described Charcot's triad of jaundice, fever with rigors and upper abdominal pain associated with gallstones, Charcot's syncope – caused by coughing, and Charcot Weiss baker syndrome of transient syncope associated with slowing of heart rate and low blood pressure to name a few.

The books illustrated that his interests lay not only in nervous diseases.

He described kidney diseases such as Bright disease, general medical conditions and was one of the first physicians to believe that diseases of not only women but also the elderly and those with chronic disease and disability were worthy of study and treatment.

For myself as an orthopaedic surgeon of course, I know the name Charcot for its association with the "Charcot Foot" and with Charcot Marie Tooth disease.

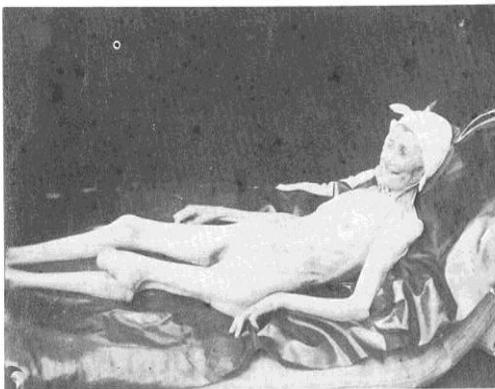
I know that all of you will have come across the confusion caused by these two diseases which both carry his name.

Charcot disease is not actually a disease but a condition – now usually due to diabetes – in which feeling is lost-including protective feeling in the joints – which then develop severe and deforming arthritis, swelling and eventually infection.

In describing neuropathic or Charcot joints, Charcot had a plentiful supply of patients not of diabetics but of women with longstanding syphilis.

His first description of the painless swollen Charcot joints of syphilis was published in 1868- and typically of the Charcot disease of syphilis it affected mostly the knees.

He continued to observe draw, photograph and record the changes over many years and 13 years later..



Figures 5 and 6
Two museological procedures of diseases.
Fig. 5: photograph excerpted from one of Charcot's clinical dossiers;
fig. 6: cast of the same "case."

In 1881 he presented a case of disease affecting the knees to the International Medical Congress of London.

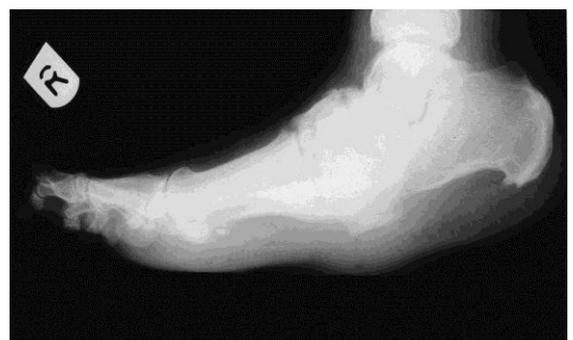
To illustrate the case he took by boat a wax model of a 60year old woman- shown here - with photographs of her bony deformities in life.

He also apparently transported her skeleton to London for the demonstration- something which I suspect modern customs officers would prevent.

Two years later, at a meeting of the French anatomical society he reported involvement of the foot for the first time.

Nowadays with increasing levels of diabetes in the community, and improved survival of diabetic patients, Charcot foot presents with increasing frequency. But in Charcot's time, diabetes was rapidly fatal, and Charcot's disease of the foot in diabetics was not in fact described for nearly 70 years.

However the association of Charcot's name survived the changes in aetiology which occurred because of medical progress.



But it is not Charcot disease but Charcot Marie Tooth disease which brings us here today.

Charcot Marie Tooth disease is of course as we have heard at this and previous meetings a mixed bunch of genetic hereditary sensory motor neuropathies.

It was first described around the time of Charcot but over a number of years and by several observers.

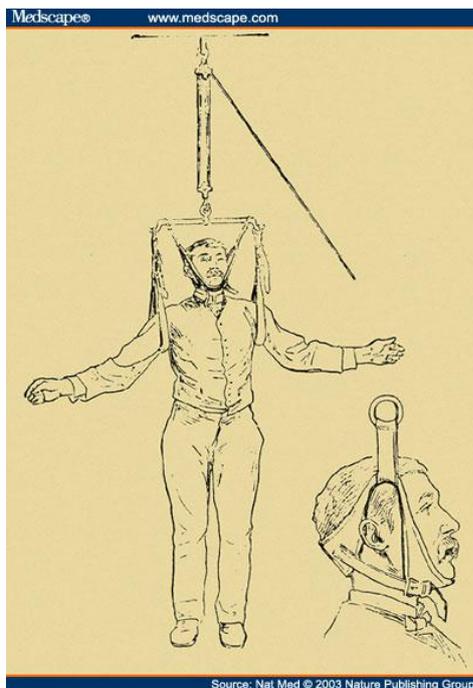
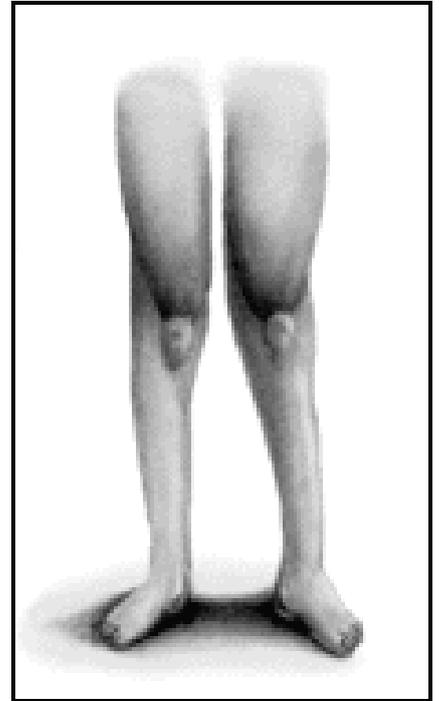
But they campaigned for the disease to bear the names of Charcot and Marie, when they described the clinical syndrome – probably of type 1a CMT in detail in 1886.

Charcot was already professor of Neurology. Marie was his student and secretary, but at the age of 33 was already a neurologist of rising reputation in his own right.

Together they described a progressive muscular atrophy of distal muscles, affecting the lower limbs first and producing the wasting later described as inverted champagne bottle appearance.

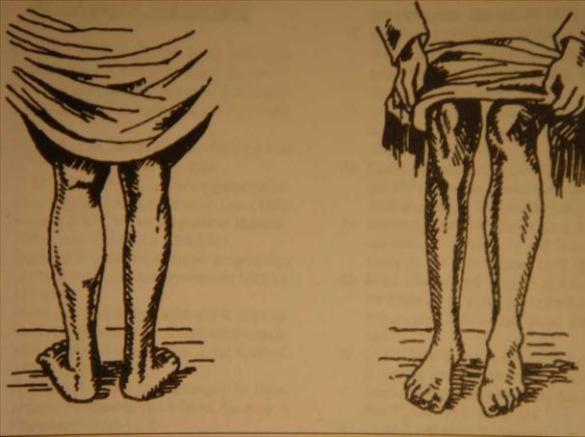
Describing in detail only 5 young patients, they noted that the disease usually began in childhood and was progressive in its weakness and the secondary deformities as shown here of high arch and inversion of the foot.

They noted that it commonly affected the siblings and forebears.



They were uncertain about the cause of the condition and wrote: “Is this a myelopathy (meaning a trapping of the nerves in the neck) or multiple peripheral neuritis – a disease of the nerves itself?” They went on to write: “The question is complicated although we prefer the hypothesis of myelopathy”.

In this one single aspect, of course they made the wrong choice, and this led to their false belief that the condition might be treated by traction to relieve the trapping of the nerves– the picture shown here is of the method they developed and favoured – which to the modern eye seems to combine futility and danger in equal proportions. Many here today should be glad that nerve entrapment is not the cause and that this has been shown to be useless as a treatment.



At the same time, Henry Tooth from Cambridge published his thesis “The Peroneal Type of Progressive Muscular Atrophy” from which this picture is redrawn. He described weakness “most often of the peroneal muscles in a disease in which heredity is a marked feature”. He also hypothesised correctly that the disorder was caused by peripheral nerve dysfunction. As a result of his contributions – and especially his correctly attributing the disease as a disease of the nerves themselves his name was soon added to those of Charcot and Marie.

It is a testament to all three of these men that the clinical and pathological descriptions of CMT barely changed in the next 130 years, until new techniques of molecular biology began to uncover the origins of the various forms of CMT .



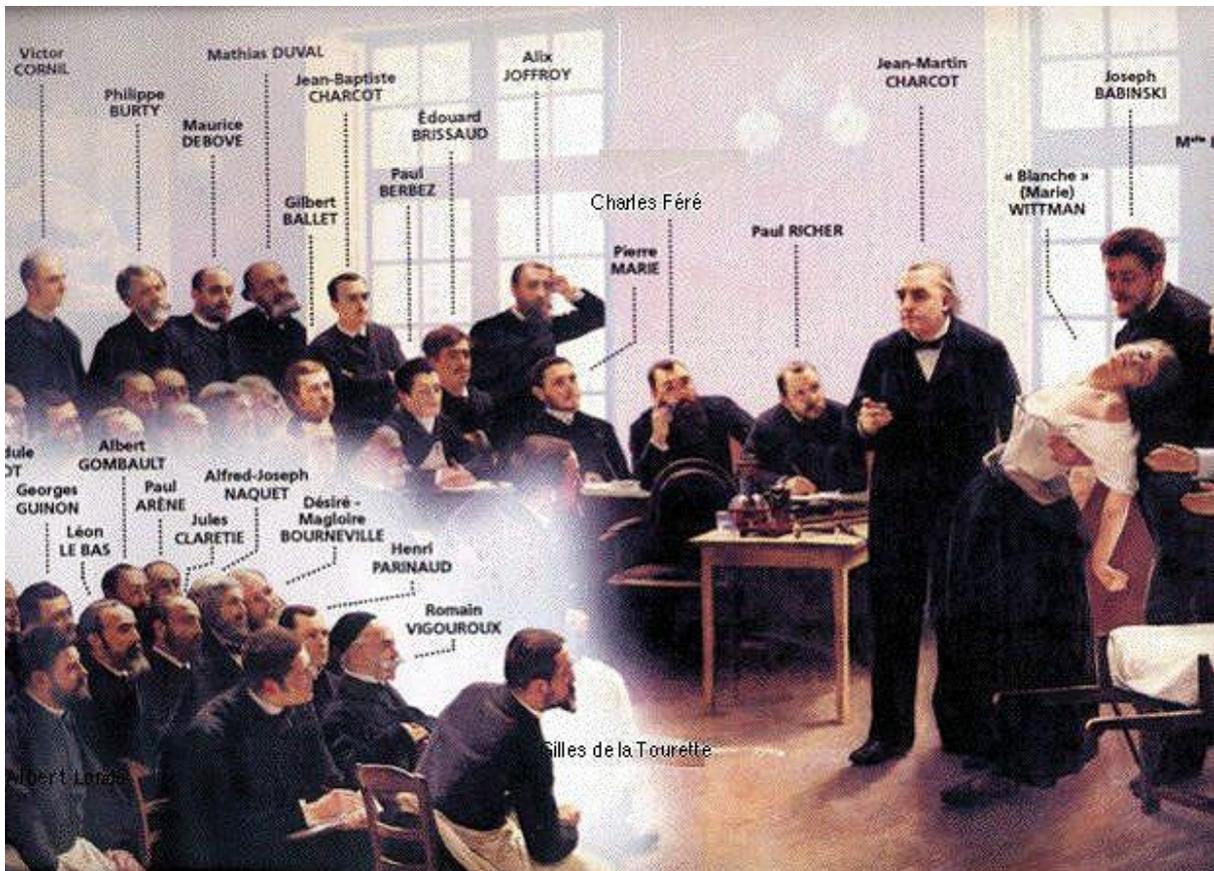
So as well as all these clinical achievements what else did he do? Well, Charcot understood the value and importance of bringing trainees into the hospital and led and encouraged teaching.

He created free teaching courses – the Friday morning rehearsed lectures.

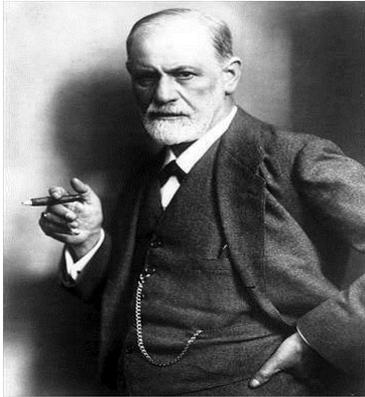
It was at these lectures that Charcot and Vulpian gave the first report on Multiple Sclerosis) on 9 May 1866, when they described three cases with careful clinical observation and autopsy findings.



A later addition was the Tuesday neurology teaching clinic for doctors, shown here in his words “describing and understanding the mechanisms of hysteria, emphasizing “the psychic determination of organic symptoms favoured by a susceptible personality.



All the participants in the painting have been identified, including Charcot himself...but you may notice other famous names amongst those present including Marie, and Charcot's son Jean Baptiste. One of the legacies he left was that he had many pupils that he encouraged and guided who went on to be famous in their own right:



Sigmund Freud is of course known to the world for his contributions to psychoanalysis and psychotherapy. He went to work for Charcot early in his career when he could not decide between medicine and the law. After 6 months working for Charcot he published his first paper on the psychosexual origins of disease jointly with Charcot and decided to make his future as a doctor.



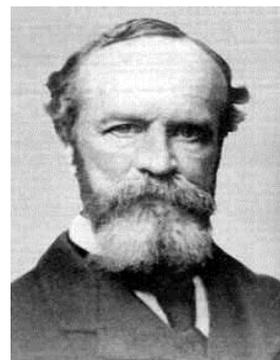
Pierre Marie described with Charcot the neurological disease that bears his name, and which brings us here today, but also wrote papers and books on neurological disease.

Joseph Babinski was a famous neurologist - familiar to all of us for his description of Babinski's sign, which all patients detest- for he described that horrible sign of scratching the sole of the foot to elicit response which most of you will have experienced.



Pierre Janet is perhaps not so well known outside France but described dissociation and traumatic memory disturbance

William James, who was the brother of the American author Henry James, came across the Atlantic to study under Charcot. He went on to publish on educational psychology, the psychology of religious experience and mysticism.





Duchenne who we will recognise for describing muscular dystrophy, but whose main research was into faradism and electrical stimulation of muscles as entertainingly applied here.

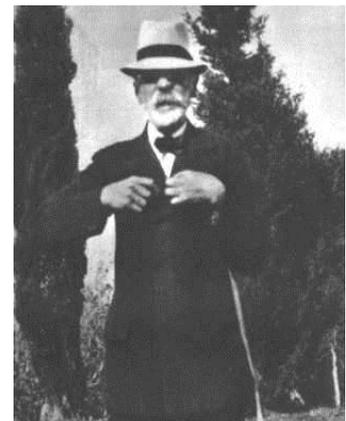
Albert Londe the early medical photographer who worked with Charcot at the Hôpital de la Salpêtrière.

You have already heard about his achievements in developing cinema.



George Gilles de la Tourette described the syndrome which bears his name with the blessing of his chief. The syndrome is applied to those people who unintentionally scatter their conversations with swear words and obscenities.

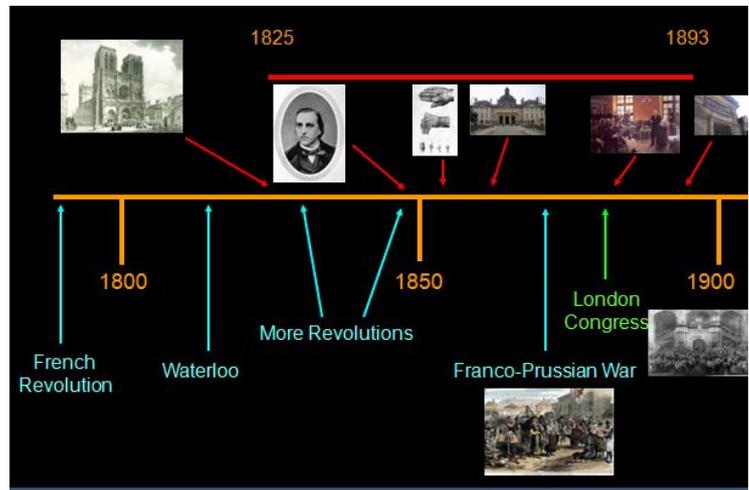
Axel Munthe went on to write his famous autobiography: The Story of San Michele.



Finally, Alfred Binet described the first scientific intelligence test, which led to the development of the modern IQ test often still referred to as the Binet tests.

An extraordinary heritage just of pupils

After achieving all this, Charcot died of a heart attack near Lake Settons in Nièvre on August 16 1893, at the age of 67. In this life he had risen from relative poverty, survived 2 revolutions and the siege of Paris.



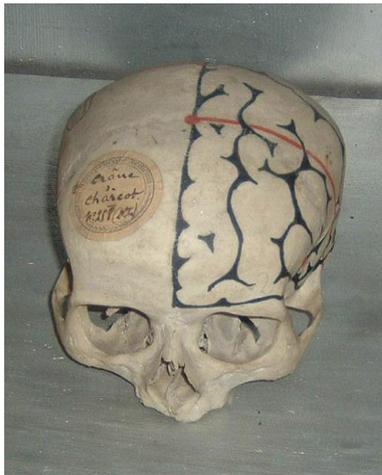
It is difficult for us to understand, but scientists of his time were the A list celebrities of the era.

Whilst alive he made was an officer of the Legion d’Honneur, vice president of the Societe de Biologie, and president of the Societe Anatomique.

In 1882 he was created Docteur d’honneur at the University of Wurzburg at its 300th anniversary, and in 1883 his greatest honour was bestowed when elected a member of the Academie de

Sciences. He was the David Beckham of his time – and like David Beckham accrued a considerable fortune from his work.





As was the accepted fashion for famous scientists in Continental Medical Schools in the eighteenth and nineteenth centuries, although not in Britain, his skull was preserved, and is now in the Museum of the Medical School in Montpellier, marked appropriately with the convolutions and areas of the brain.



So I hope that in a relatively short time I have taught you something about a very remarkable man, who makes up the C of CMT.

He was inquisitive and innovative.

He recorded with accuracy and taught with enthusiasm, leading and encouraging his pupils.

He travelled, but still returned home to a family of which he was proud.

He led others to believe in the rights of psychiatric patients, women and the elderly.

He abhorred and campaigned against unnecessary suffering caused to people or animals – and hated the English as a nation of foxhunters and animal abusers.

Working in ridiculously difficult times he did enormous good.

“He was a personality of high intellectual and moral culture, a professor of exceptional value, a creator in the domain of science”
Georges Guillain (1876 – 1961)

And this would be the end of the presentation but for one fascinating follow on.

And here may I ask whether any of you have been on a cruise to the Antarctic? I haven't!



We have already seen a picture of his son, Jean-Baptiste Charcot (1867-1936) who practised medicine. he inherited his Father's fortune in 1893. Well as soon as he got the money, he immediately stopped being a doctor , used the fortune to buy a boat and went on to become one of the great polar explorers.

He was known as Commandant Charcot, and was known as "The Gentleman of the Pole" according to Captain Robert Scott.

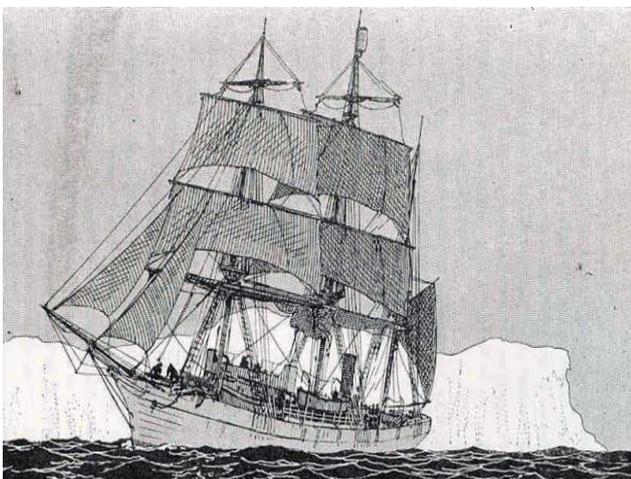


He is remembered for the French Arctic and Antarctic exploration aboard "Le Français" and "Le Pourquoi-Pas" from 1893 to 1910, He later served and was decorated as commander in the British Navy during the great war.

This is his first ship, "Le Français" .

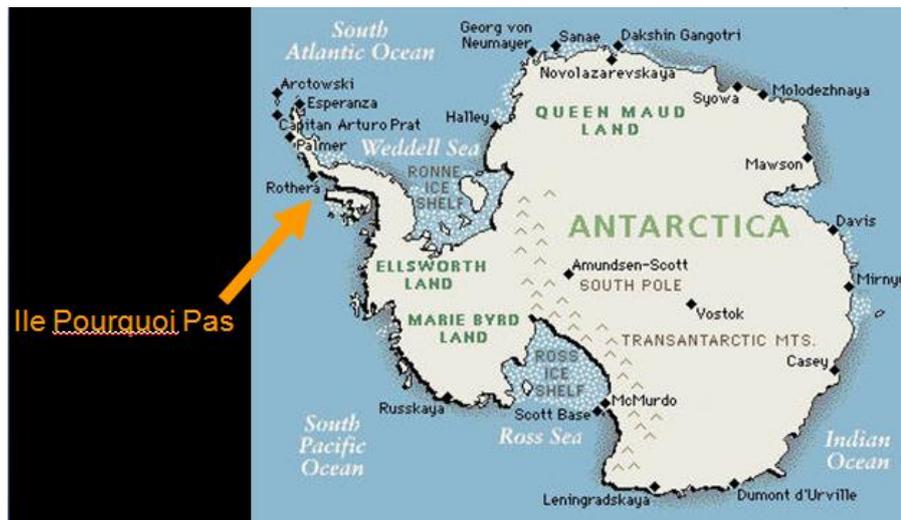


Later he sailed in the "Pourquoi Pas?" which was named after a model boat that he had sailed with his father on a lake as a child. In it he was part of the international group that searched for Shackleton.



In the Antarctic, his explorations were mainly around the Peninsula, which is now a destination for tourist cruises.

There is still an island named after his boat just South of the Antarctic Circle.



- Charcot Island
- Cape Charcot
- Port Charcot
- Charcot Cove
- Charcot Bay
- Charcot Fan

Several Antarctic landmarks are named after Jean-Baptiste or by Jean-Baptiste after his Father, you can see them listed here and those of you who may have travelled to the Arctic or Antarctica may have wondered how these places have got their names.

Well I hope that now all of you whether you ever travel to Antarctica or not will think of Charcot father and son as two remarkable people and not just the C of CMT.

