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Review Article

From frogs' legs to pieds-noirs and beyond: some aspects of cochlear implantation

GRAHAM FRASER MEMORIAL LECTURE 2002

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Abstract

The 2002 Graham Fraser Memorial Lecture deals first with the French origins of cochlear implantation in Paris in the 1950s and the role of André Djourno and Charles Eyriès. Following this work in Paris Dr William House in Los Angeles continued work on cochlear implants and, subsequently, experimental implant programmes were started in California, Paris, Vienna and Melbourne.

The next section of this lecture covers the experimental work of Galvani in establishing the role of electricity in physiology. The results of his first experiments were published in 1791, the year that Mondini produced the first account of a cochlear malformation in a congenitally deaf child.

At around the same time sign language for congenitally deaf children was being developed for the first time in Paris by Epée and the first disputes occurred between oralists and those who promoted signing for the education of congenitally deaf children.

In a present day cochlear implant programme good results from implanting congenitally deaf children at an early age and implanting adults who have become profoundly deaf are now taken for granted. We do have much to learn, however, from more complex implant candidates and some examples of such candidates are presented.

Lastly, looking to the future, the use of PET scanning to try and gain information about how the brain handles the information provided to it by a cochlear implant is described.

Key words: History of Medicine, 18th Cent; Cochlear Implants; Tomography; Emmission-Computed; Deafness; Sign Language

I am grateful to the trustees of the Graham Fraser Foundation for kindly inviting me to give this eighth Graham Fraser memorial lecture. In this lecture, I will start by covering three areas of past work that are relevant to the present-day clinical practice of cochlear implantation. Then I would like to describe a few current areas of interest in this field. Finally, I will mention some possible future directions of implant design.

Firstly, I would like to go back to Paris in the 1950s, with a story that involves two pieds-noirs. 'pieds-noirs' literally means 'black feet'. Pieds-noirs are French people who were born in the previous French colonies in North Africa, especially Algeria. Their feet are considered 'black' from treading the soil of Africa, although their families were originally from mainland France. It is a description of which pieds-noirs themselves are proud. After the Algerian war of independence many pieds-noirs moved to *l'Hexagon:* mainland France.

André Djourno was born in Algeria in 1904. He trained as a doctor in Algeria, and moved to Paris in 1935. He eventually became professor in the medical faculty of Paris University. His main field was innovative research in physiology. For example, when the French Electricity Company changed its current from 120v to 240v, Djourno was asked to look at the risks of electrocution. From there he became interested in resuscitation and must have been one of the first people to build a cardiac defibrillator.

He also looked at other applications of electricity in the field of physiology. He buried small coils in experimental animals. These coils consisted of a centre core of iron, with wire wound tightly around it. One or both ends of the wire were put in contact

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with the nerve or muscle to be stimulated, the skin was closed and allowed to heal. Using the principle of electrical induction, another coil, connected to an electric current, was put on the outside of the skin, over the buried coil. Current flowing through this external induction coil produced an electromagnetic field which passed through the skin and soft tissues to induce a matching current in the implanted coil. One application of this technique was to stimulate the phrenic nerve to activate the muscle of the diaphragm and so provide a simple, cheap and portable replacement for the extracorporal method of ventilation, the 'iron lung', then used to provide respiration for people paralysed by polio. Djourno implanted these coils into frogs and rabbits,[1] and collected experimental data on the ability of the body to tolerate the presence of the implants, and the ability of the nerves to withstand long-term electrical stimulation.

He also had the idea of using this technique to stimulate the cochlear nerve in deaf patients.[2] If, as a French academic, you needed to establish precedence for an invention or discovery before full publication, there was a system of depositing your notes in a *pli cachété* (sealed envelope) with the Academy of Science. Djourno later described having done this in 1953 with his concept of a cochlear prosthesis.[3]

Four years later, in 1957, Djourno was approached by Charles Eyriès, another pied-noir, from a French Algerian family who had settled in Paris. Eyriès had the reputation of being an excellent otologist. He had been sent a patient with complications related to cholesteatoma in both middle ears. Ten years earlier this man had had left-sided radical mastoid surgery, leaving him with no hearing in the left ear and a left-sided facial paralysis from damage to the facial nerve. Around 1957 he had a similar procedure on his right ear, sadly with the same result.

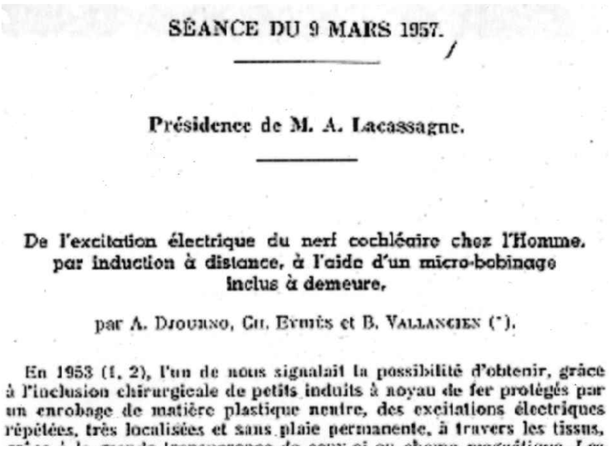
The patient, a man in his 50s, was thus left with no hearing in either ear and a completely paralysed, immobile face. What Eyriès did first was to explore the right ear under local anaesthetic in an attempt to repair the facial nerve. This turned out to be technically impossible, because of what Eyriès described as the abnormal slenderness of the nerves normally used for grafting the facial nerve. Eyriès decided that he would make another attempt later using foetal sciatic nerve.

During this first operation, Eyriès had used electrical diathermy while exploring the ear. Whenever the diathermy was used, the patient (the procedure was under local anaesthetic) said that he received a sensation of hearing. Later, the patient, who was an engineer, drew Eyriès' attention to the fact that the diathermy appeared to produce a sensation of sound, and asked whether it might be possible to find a technique to help his hearing. Eyriès knew of Djourno's work through colleagues, and approached him. Djourno agreed to try and help. With his assistant, Danièle Kayser he constructed an appropriate coil, which was embedded in epoxy resin and sterilized.

Figure 1 is a copy of the title page of the first published article describing cochlear implantation. It is from *Comptes Rendus* (proceedings) *de la Societé de Biologie* in Paris on 9 March 1957.[4] The article is by Djourno, Eyriès and Vallencien, and mentions the technical assistance of Danièle Kayser. The article mentions Djourno's earlier animal experiments, and then goes on to say: 'a patient who had suffered extensive damage to both ears asked if it might be possible to get rid, even partly, of the total deafness with which he was afflicted. This desire was so strong that, after warning him of the likelihood of failure, we agreed to implant a coil during an operation to graft his facial nerve on February 25. After inserting a 5 cm facial nerve graft, we found such extensive destruction that at first we hesitated to implant the coil. Eventually, however, we went ahead; partly for obvious psychological reasons and also because we were able to identify a small stump of nerve only a few millimetres long, but accessible enough to place the electrode in contact with it without putting the patient at risk' (presumably this meant without opening the internal auditory meatus).

Figure 2 shows three coils. The central one with two ends was implanted; it is 2.5 cm long and 3.5 mm thick, with two terminals of stainless steel wire. 'One wire was insulated with polythene as far as its tip and placed in contact with the rather frayed stump of nerve. The other end, not insulated, was buried with the coil itself in the temporalis muscle'. Figure 3 is a plain X-ray of the device in place. The coil and the indifferent electrode can be seen, but the active electrode is not easy to identify.

The first tests were done three days after the operation, rather sooner than the six weeks post-operatively that is current practice. The external coil (the upper one in Figure 2) was connected to an amplifier that Djourno had previously built to stimulate the phrenic nerve in rabbits. This amplifier gave 15–20 bursts a minute of a 100 Hz alternating current. When the external coil was held at a distance from his head the patient said that he could hear a



SÉANCE DU 9 MARS 1957.

Présidence de M. A. Lacassagne.

De l'excitation électrique du nerf cochléaire chez l'Homme, par induction à distance, à l'aide d'un micro-bobinage inclus à demeure,

par A. Djourno, Ch. Eyriès et B. Vallancien (*).

En 1953 (1, 2), l'un de nous signalait la possibilité d'obtenir, grâce à l'inclusion chirurgicale de petits induits à noyau de fer protégés par un enrobage de matière plastique neutre, des excitations électriques répétées, très localisées et sans plaie permanente, à travers les tissus,

Fig. 1

Reprinted from *Comptes Rendus de la societé de Biologie*, March 9th 1957

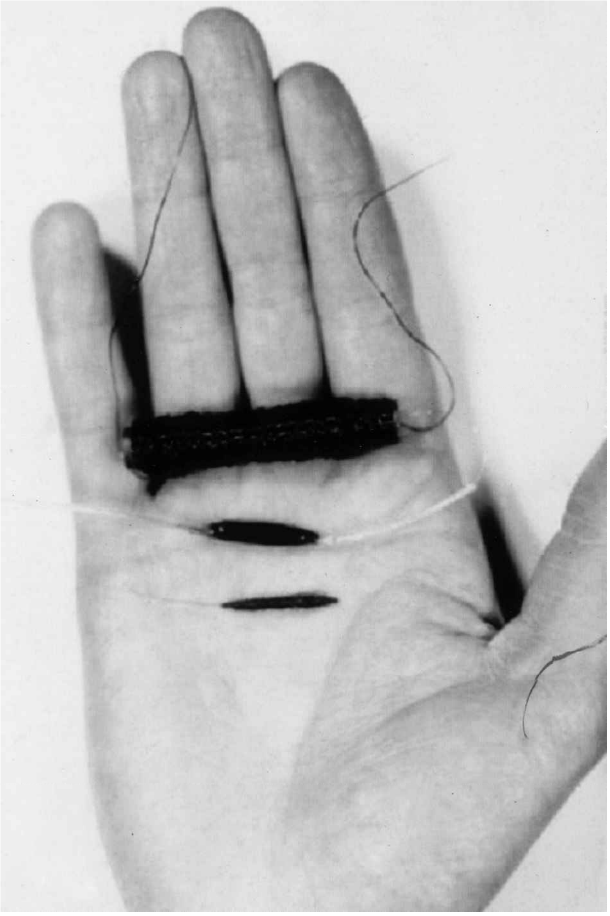


FIG. 2
Three of Djourno's coils. The upper one is the stimulating, external coil. The middle one is the bipolar receiving coil that was implanted. The lowest one is monopolar and was not used.

sound like crickets. As the external coil was brought closer to his head, the sound became louder and he described it as being like a squeaky wheel.

This report[4] was presented only eight days after the operation. Five months later there was a second report,[3] this time in a different journal, *La Press Medicale*, with details of more tests. They found that the patient could distinguish between different stimulation frequencies up to 1 kHz. Above 1 kHz, all frequencies sounded the same: shrill but not unpleasant. The induction coil was connected to a microphone and although the patient was not able to understand speech fully he was, after a time, able to identify words from a small closed-set group of words, including *maman, papa* and *'allo*. Sometimes, he could correctly guess other words, such as *bravo*.

Unfortunately, after a month, the wire that formed the indifferent electrode fractured, and the device stopped working. Eyriès re-explored the ear, and implanted a new coil. This functioned just as well as the previous one. The patient delighted in being able to hear. He practiced using the device on his own, talking to himself, and loved listening to conversations taking place around him and even doors opening and closing. He recovered some facial movement on the operated side. There was no unwanted stimulation of the facial nerve when the device was in use, nor was there any disturbance of balance. Sadly, the second device broke down, probably for the same reason that the first device had stopped working. In Eyriès' later account in 1979,[5] he said that he decided not to risk a third operation because of the patient's poor state of health. In fact, the patient died of a heart attack 20 months after the first operation.

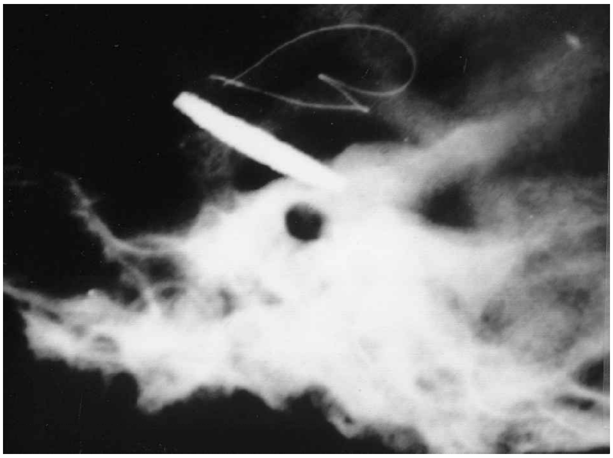


FIG. 3
Plain lateral skull X-ray, with the implanted coil in place.

At this stage, a disagreement arose between Djourno and Eyriès about the potential commercial value of the implant. It seems that Djourno had strong views on this. He apparently felt that, as a matter of principle, scientific advances ought to remain totally within the public domain, and he refused to consider patenting the device. He did construct another device to be used by a different surgeon, Roger Maspetiol. The choice of the patient was less good. She was a young Vietnamese woman deafened by streptomycin prescribed for tuberculosis.[6,7] The device did give her a sensation of hearing, but she seems not to have been keen to receive the implant in the first place. Six months after the operation she returned to Vietnam. She is recorded as having described electrical stimulation as sounding like the *hou hou* sound of the wind.[7] In 1958 Djourno also described using promontory stimulation in this patient, with a transtympanic needle to establish the integrity of the cochlear nerve.[6] This was the first description of the use of a transtympanic needle electrode, predating Portmann LeBert and Aran,[8] by nine years; it also predates House and Brackmann[9] by 16 years as the first description of using this electrode for electrical promontory stimulation.

Djourno carried on with his experimental work.[10,11] He had recognized the need for better frequency discrimination, separating sounds of different frequencies and sending them to separate electrodes. He built a machine that was both able to identify different speech frequencies and to perform real time frequency analysis of speech, including vowels, using an oscilloscope. Different frequencies were channelled to different electrodes: all the features of a prototype multi-channel cochlear implant. There were rabbit and frog experiments in which a low-frequency sound, for example, was used to stimulate the left leg of the animal, and a high-frequency sound the right leg.

By 1959, Djourno had, therefore dealt successfully with much of the theory and practice of a multi-channel cochlear implant and also pioneered the use of a transtympanic needle and transtympanic promontory stimulation, on his own and starting from scratch. He and his team had 12 publications, two patients and a great deal of experimental data. At this stage, however, he felt that he needed more funds: he particularly needed to employ an engineer. A grant application was turned down and, as mentioned earlier, he was unwilling to consider any commercial involvement. He therefore simply called a halt to this work and moved to something else. His view, apparently, was that he had done what he could, putting the results in the public domain where they were available for other workers and that was that.

Djourno died in 1996, at the age of 92 and is buried with his wife in the Monparnasse cemetery. Eyriès died around the same time, and is buried in the village of Charly, near Bourges, south of Paris. Mlle Kayser still lives in Paris. When Djourno retired, she moved into the field of electric response audiometry. I am grateful to her for many of the details and background contained in the above account. I should also acknowledge the help and advice from Phillip Seitz, a historian who researched the French origins of cochlear implantation and interviewed both Djourno and Eyriès and Mlle. Kayser while working for the American Academy of Otorhinolaryngology Foundation.[12]

In 1961, four years after Djourno's first paper, Dr William House, in Los Angeles, heard of Djourno's paper and had it translated into English. With the help of an engineer some implants were made and implanted. However, the work stopped, once again, it seems, because of a disagreement about commercial applications. It was only in 1972, 15 years after Djourno and Eyriès' first paper, that House restarted a single channel cochlear implant programme with an engineer called Jack Urban. Parallel work took place in California, Paris, Vienna and Melbourne. Chouard, who developed the implant programme in Paris, knew Djourno and had also worked with Eyriès during his training. It is interesting to speculate what might have been the present state of development of cochlear implants if Djourno had received his grant and continued his work during the 15–20 years during which there was little published activity in the field of implantation.

That deals with the second part of my title, the pieds-noirs. For the first part of my title, 'frogs legs', we need to go back in time to 1791. This was the year Mozart died in Vienna, on 5 December. At the same time, about 300 miles to the south-west of Vienna, in Bologna, in Italy, the foundations both of neurophysiology and of the harnessing of electricity were being laid.

Figure 4 shows an extract from the quarterly *Commentarii*, or Records, of the Bologna Institute and Academy of Science and Art. It is from an article by Luigi Galvani.[13] On the left is a portrait of Galvani and on the right of his wife, Lucia Galeazzi, the daughter of a senior professor in Bologna. Lucia Galleazzi worked with her husband in his frog experiments, and it is good to see her mentioned in this first article by Galvani. A portion of the text in the left-hand column is expanded in the right column and reads 'Assistando uno, la moglie o altero un dito al conduttore' 'When the wife or someone puts their finger near the electric plate there is a spark'.

Figure 5, which happened to be displayed in the hall of the Royal Society of Medicine on the day when this lecture took place, shows the experiment in process. Static electricity, which can damage a cochlear implant if the scalp directly over the implant touches the door arch of a car or the plastic of a playground slide, was well recognized in 1791. People knew a great deal about lightning, electric fish and the Leyden jar in which static electricity could be stored. There were electroscopes that were able to measure the size of an electrical charge.

Galvani's first experiment, shown mainly on the left of the picture, includes the large wheel used to generate a charge of static electricity. Also on the left of the picture is a rather large-scale frog's leg preparation. You can see the two legs, and spinal cord. A charge of static electricity was built up using the wheel. When someone touched the charged apparatus (indicated by the disembodied hand pointing downwards, near the top of the centre of the picture), there was a spark as the electricity

FIG. 4

Luigi Galvani, Lucia Galleazi and the manuscript of Galvani's first article. (Reprinted with kind permission of Professor Marco Piccolino.)

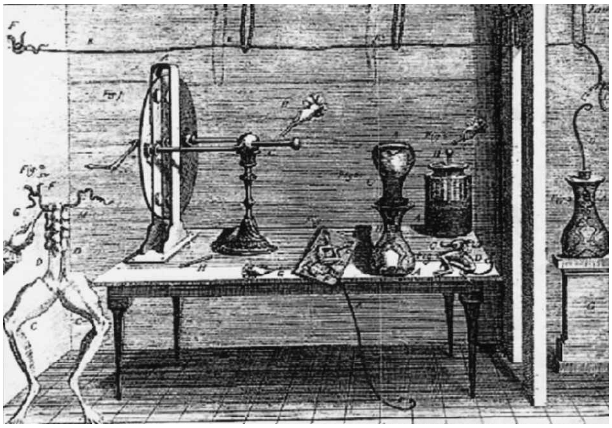


FIG. 5

Plate 1 of the Galvani's article in the 1791 Commentarii of the Bologna Institute and Academy of Science and Art. The prepared frog on the left of the picture and the static electricity-generating wheel are intended to demonstrate the 'spark' experiment.

stored in the device discharged itself into the atmosphere. It seems that this may have happened accidentally while Galvani was holding a metal scalpel with an insulated handle against the frog's crural nerve (the hand on the left). The same instant that the spark was generated by discharging the static electricity machine, the frog's legs contracted. It was already well known that static electricity could stimulate muscles. However, in this case, it was the quite small but very abrupt discharge of electricity into the atmosphere that appears to have been picked up by the insulated metal scalpel, passed to the spinal cord and triggered the movement of the frog's legs.

Galvani's next experiments led to two quite different sets of conclusions, both relevant to the later development of cochlear implants. Figure 6 shows the frog's leg preparation attached to a long wire suspended across the roof of a building. Galvani wanted to see whether the natural discharge of static electricity into the atmosphere during a thunderstorm could be collected by the metal wire and would stimulate the frog's legs. The experiment worked: with every flash of lightening, the legs moved. Galvani next wondered whether there was enough electricity in the atmosphere in more normal conditions to produce a similar effect. He used a copper hook to suspend the frog's leg preparation from the metal rail of a balcony. This seems to have had no effect, but, possibly while starting to unhook the frog's legs, one of them accidentally touched the metal of the balcony and the leg twitched.

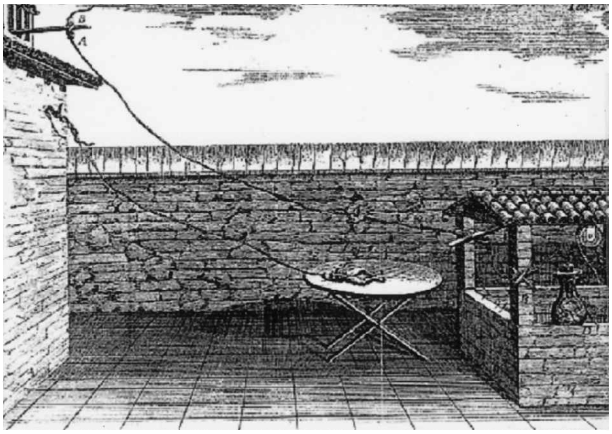


FIG. 6

Plate II of the 1791 Commentarii article by Galvani. The roof-top experiment with atmospheric electricity during a thunderstorm.

Galvani's conclusion was that the energy that causes muscles to move was not the older and rather vague concept of 'animal spirits' but 'animal electricity', stored in minute quantities in the muscles fibres of all living creatures.[14] He established that there was a non-linear relationship between the strength of the stimulus and the size of the contraction, and that the contractions fatigued with repeated stimuli, then recovered. Galvani's insight was correct, although it took many years and conclusively the work of Hodgkin and others[15] at University College, London in the 1930s, 40s and 50s before technology caught up with Galvani's insight and proved him correct.

Meanwhile, Alessandro Volta, a younger scientist working in Pavia, repeated Galvani's experiments and suspected that the last experiment showed something else. His hypothesis was that the wet frog's legs acted as an electrical conductor between two different metals, the copper hook and the iron balcony rail, completing a circuit between them. He felt that the contraction of the frog's legs was therefore nothing but a way of detecting the generation of an electric current flowing between the two different metals. In this way, the frog acted as a much more sensitive indicator of electricity than even the most sensitive electroscope. Volta eventually increased the strength of the electric current by piling up a sort of club sandwich of alternate zinc and silver plates, each separated by a cloth soaked in salt water, and so constructed the first battery.[14]

Politics then came in. Napoleon invaded Italy. Galvani, the older man, did not welcome this; he lost his professional chair and died in 1798. Volta, on the contrary, was fêted by Napoleon, demonstrated his battery at the Royal Society in London and became famous. Galvani's hypothesis was largely ignored, until von Helmoltz[16] and Bernstein[17] reactivated the study of electro-physiology in the mid-1800s, Hodg-

OPUSCULA,

CAROLI MUNDINI

Anatomica ſurdi nati ſectio.

FIG. 7

The title page of Mondini's first description of a cochlear malformation.

kin, Huxley and Katz[15] did their giant squid axon experiments, Dawson,[18] Davis,[19] Portmann and Aran[8] worked on the electro-physiology of hearing and Djourno built his first cochlear implant. Eventually, therefore, Galvani's balcony experiment proved his hypothesis correct: this crucial experiment turns out to be the starting point both for the science of neurophysiology and for the practical use of electricity by mankind. It is interesting that the first of these was intended by Galvani, but accepted only after considerable delay, while the second was unintended, nearly immediate, and resulted from a rival interpretation by a younger scientist.

In passing, here is a coincidence: Galvani's first paper was published in the proceedings of the Bologna academy of science and art, vol 7, pages 363–418.[13] On the very next page, p 419 (Figure 8) in the same issue of this quarterly journal, is the first description[20] by Carlo Mondini of a congenital malformation of the cochlea.

Mondini's account of what is now known as the Mondini malformation: a cochlea with only one and a half turns, rather than two and a half, and a wide vestibular aqueduct, comes from the post mortem study of the skull of a congenitally deaf child of nine who died after a road accident (he failed to hear the approach of a speeding wagon). The Mondini malformation is of great interest to cochlear implant surgeons, since it, and other forms of congenital cochlear malformation, can lead to difficulties in the insertion of cochlear implant electrodes and to 'gushers' of cerebrospinal fluid (CSF), when the malformation is associated with an open connection between the perilymph in the vestibule of the cochlea and CSF in the internal auditory meatus.

FIG. 8

Abbé Charles Michel de l'Epée (1712–1789).

Let us stay in the same year, 1791, when Galvani and Mondini both published in the quarterly Bologna journal, and move to Paris, leaving behind the scientific roots of cochlear implants to look at what was to become one of their most successful applications: the treatment of congenitally deaf children.

The Bastille had been stormed two years earlier, on 14 July 1789, and the French Revolution was getting into its stride. Republicanism, regicide and the invention of the guillotine and the metric system were all in the air. In 1791, the *Marseillaise* was heard on the streets of Paris for the first time, sung by gangs of Fédèrés from Marseilles. Two years earlier, the Abbé Epée (Figure 8) had died.

When he was 48 Epée found himself handed the job of giving religious instruction to congenitally deaf twin sisters. Over the next 30 years, he developed a formal system of signing to communicate with the deaf. Originally, it seems, this was an adaptation of the childhood signs he and his schoolmates had used to communicate behind their teachers' backs. Using this new 'sign language' he built up a famous school for deaf mutes in the Rues des Moulins, in Paris. On Epée's death, the State agreed to take over the school and fund it. One of his previous assistants, Abbé Roche-Amboise Sicard (Figure 9), was recalled from Bordeaux, and appointed head of the school. So, in 1791, the National Institution for Deaf Mutes was founded. Unfortunately, as a priest, Sicard was a potential target for the Revolution. The following August he was rounded up and, in spite of a spirited defence by a delegation from his school, sent to the Abbaye Prison, along with 23 other priests.

Not long afterwards, a mob stirred up by Danton stormed the prison, found the group of 24 priests and hacked 19 of them to death with a variety of instruments, including a butcher's saw. Sicard was one of the five survivors, recognized by a butcher, possibly one of his local tradesman.

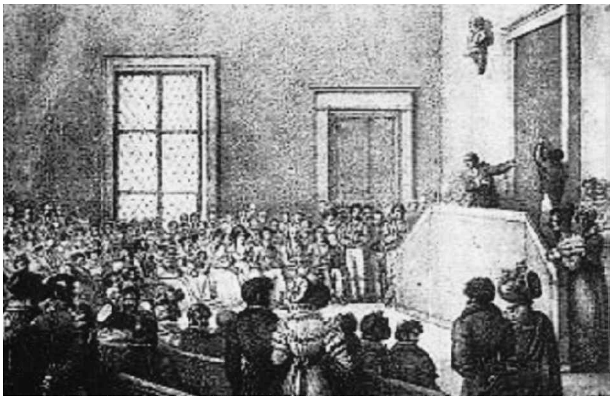
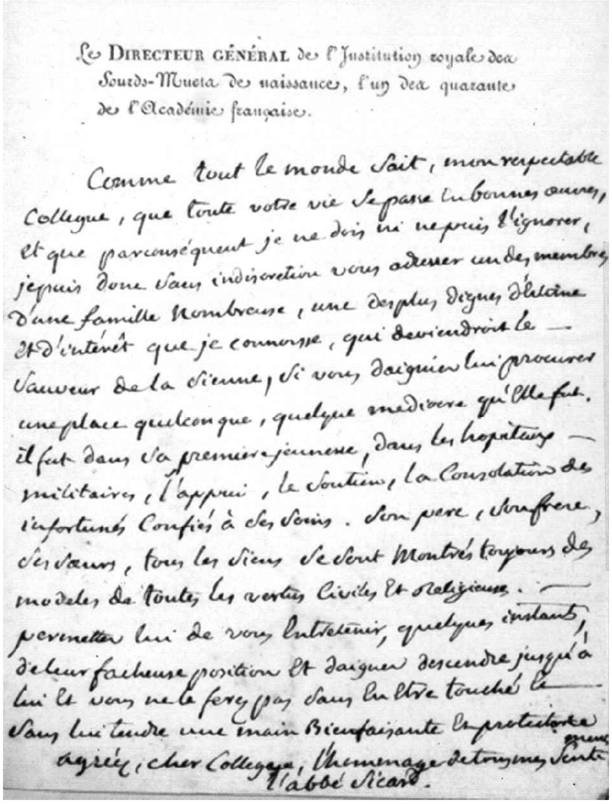


FIG. 9

A public lecture at the Institution National des Sourds-Muets in Paris, c 1810, showing Sicard addressing the audience and Massiue, his assistant, at the blackboard, chalk in hand.



Le Directeur Général de l'Institution royale des
Sourds-Muets de naissance, l'un des quarante
de l'Académie française.

Comme tout le monde sait, mon respectable
Collègue, que toute votre vie se passe en bonnes œuvres,
et que par conséquent je ne dois ni ne puis l'ignorer,
je puis donc sans indiscrétion vous adresser un des membres
d'une famille nombreuse, une des plus dignes d'estime
et d'intérêt que je connoisse, qui deviendroit le
sauveur de la sienne, si vous daigniez lui procurer
une place quelconque, quelque médiocre qu'elle fût.
il fut dans sa première jeunesse, dans les hôpitaux
militaires, l'appui, le soutien, la consolation des
infortunés confiés à ses soins. son père, son frère,
ses sœurs, tous les siens se sont montrés toujours des
modèles de toutes les vertus civiles et religieuses.
permettez lui de vous entretenir, quelques instants,
de leur facheuse position et daignez descendre jusqu'à
lui et vous ne le ferez pas sans en être touché et
sans lui tendre une main bienfaisante et protectrice
agréez, cher Collègue, l'hommage de tous mes sentimens
l'abbé Sicard.

FIG. 10

Letter by Sicard, dated 1818, found attached to the title page of an English translation of his work. Property of the RNID library, Royal National Throat, Nose and Ear Hospital, London.

Looking through the historical collection of the Royal National Institute of Deaf People, three weeks ago, I opened the English translation of Sicard's book on deaf education, and was surprised to find a letter (Figure 10), stuck to the title page.

Sicard wrote it in 1818 (notice that the headed paper is of the **Royal** Institution for Deaf Mutes), after a visit to England. It is quite mundane: a reference asking an English friend to find a job, however lowly, for a young doctor from a large but worthy family who had fallen on hard times. It would not have been written if the massacre described above had run its full course.

In 1791, we find the origins of a long-running controversy in deaf education that led, among other things, to a demonstration outside the Royal Society of Medicine at the time of the Graham Fraser lecture two years ago. In 1791 a young man called Jean-Marc Gaspard Itard (Figure 11), decided to dodge the draft. He had done some commercial studies but was not at all keen on doing his military service. He asked his uncle, the Abbé Itard, to find him a job as a doctor. It should be mentioned that he had not so far opened a medical textbook or set foot in a hospital. This did not prevent him being given the job of surgeon, third class, at the military hospital at Toulon. Here, he was able to attend lectures given by a famous surgeon called Larrey, who pioneered a field ambulance system and early amputation as a life-saving treatment for severe limb injuries, became Napoleon's physician, and whose life was evidently spared on the orders of the Duke of Wellington while he was treating the wounded on the field of Waterloo. (Rue Larrey happens to the address of the family home of Mlle Kayser in Paris, where she lived while acting as assistant to Djourno).

FIG. 11

Jean-Marc Gaspard Itard (1774–1838).

Itard left the army medical service as soon as he decently could, went to Paris and was appointed surgeon to the same National School for Deaf Mutes where Sicard was the Principal. Sicard is perhaps now best known for trying to teach Victor, the wild boy or wolf child, found in the woods of Aveyron, to speak, without great success. Unlike the teachers, Epée and Sicard, Itard, the doctor, seems to have been an advocate of oral education for the deaf. This difference of opinion is still a flashpoint in deaf education to which the advent of cochlear implants has brought a new perspective.

It is easy to forget the impact of powerful electric hearing aids on children with moderate or even moderate-to-severe congenital deafness in the 1940s and 1950s. Many such children, without access to speech at normal conversational levels, failed to develop the understanding of speech and so never learnt to speak. Hearing aids gave such children access to sound and so the potential to understand and produce speech. It was optimistically expected that this effect would be extrapolated to all deaf children, even those with with profound deafness; promotional literature from the National Deaf Children's Society at that time[21] reflects this optimism. Unfortunately, as an EEC report published in 1979[22] confirmed, this assumption was generally incorrect.

A cohort of children born in the European Community countries in the year 1969 were studied at the age of eight years. Of about four million children born in 1969 there were approximately 4000 with hearing loss of 50 dBHL or worse in their better ear, for frequencies averaged between 500 and 2000 Hz. Thirty-three per cent of these children had severe-to-profound deafness, with average thresholds of 100 dB or worse in their better ear and 33 per cent, using their hearing aids, could not hear anything quieter than a loud shout at a distance of three metres. The most telling statistic was that only 46 per cent of these 4000 children had speech that could be understood by people outside their own close family. This can be seen as a clear demonstration of the failure, in practice, of hearing aids to allow the most severely deaf to acquire speech.

Since the mid 1980s, cochlear implants have been shown to have an impact on profoundly congenitally deaf young children comparable to that produced by hearing aids in the moderately and moderately to severely deaf two generations earlier. There are, however, exceptions and although it is clear[23] that a majority of congenitally deaf children implanted before the age of four years will develop normal speech and understanding of speech and may attend mainstream school, this is not always the case. It is therefore one of the main duties of a vigilant paediatric cochlear implant team to avoid repeating the mistakes of our predecessors, to identify these exceptions to the rule and make appropriate recommendations and provision for them.

Having brought us to the present day I would like to deal with one or two aspects of current cochlear implant practice. Oliver Wendell Holmes said that hard cases make bad law. This may be true for law, but not medicine, where lessons learned from difficult cases can be more useful than the continued study of routine success. Some of the hardest decisions of our own cochlear implant team have been in selecting patients, particularly those who do not fit the usual criteria for implantation. One of the more difficult problems, when confronted with what we describe as a ‘non-traditional’ cochlear implant candidate, is in knowing whether we were correct in turning someone away without an implant. If we decide to give such a borderline candidate an implant, he or she may derive a great deal of benefit from it and it may improve their quality of life. On the other hand, it is also possible that they may get no benefit and, after a relatively short time, stop using the implant altogether. In either case, as we follow the patient’s progress, we will get to know whether our decision was right or wrong. However, for the people that are turned down, whether adults or children, we will never really know if we might have committed a sin of omission, so to speak, and were wrong not to offer an implant.

I will briefly describe two examples of non-traditional candidacy. A young woman joined her family in London from abroad. She is congenitally profoundly deaf and has a little, extremely unclear, speech, although she has continued to use hearing aids. She is unmarried and has a job in her own country. At her first consultation, her family interpreted for her and explained how enthusiastic she was about having an implant. They agreed that she quite understood that she could only derive limited benefit from an implant, but was very keen to have one in spite of this. An early observation of our audiologists was that the patient responded quite briskly to the sounds presented during a hearing test while she was using her hearing aids. Then, after what sounded like a sharp reminder from a relative, these responses tailed off and did not reappear. An attempt at psychological assessment with a male member of our team of psychologists left us no wiser, so a second appointment was made with a female psychologist. On this occasion, the team arranged for a professional interpreter to be available, in the absence of her own family. This time, the patient broke down and wept. She explained that the last thing she wanted was an implant and what she really wanted was to go back to her home and resume her job. She also told us that the family had felt it would be easier for them to find her a husband if she had an implant. However, she did not really want one of these either. Our team also agreed that she would receive little benefit from an implant. The situation needed extremely tactful handling, but the patient was not offered an implant.

In contrast, we have 10 teenage or young adult patients to whom we gave implants although they were by no means traditional implant candidates. They had mostly lost their hearing in the first two years of life, and had been profoundly deaf since then. In this situation, we would certainly not expect a cochlear implant greatly to improve the quality of what speech they had, or to allow them to understand speech more easily. On the other hand, we did admit to them that an implant could give them some, rather modest, benefits. They might be made more aware of when their companions were speaking, and so not interrupt them. They might be able to monitor the loudness of their own voices and, with the aid of speech therapy, gain a little more clarity in their speech.

After extremely thorough counselling, these young people have been given implants and, so far, they have continued to use them and our cautious predictions have been slightly exceeded. Of course, the ‘performance’ of these people in terms of clear, fluent speech and good understanding of speech, falls a great deal short of what we would expect from conventional cochlear implant candidates, whether adults who have lost their hearing or congenitally deaf children, implanted in the first four years of life. However, when this group of young people was tested to measure changes in their quality of life following implantation, they turned out to be getting an improvement in life quality equivalent to that recorded by the best of our ‘traditional’ adult cochlear implant patients: those who had become deaf in adult life and whose ability to hear had been restored by cochlear implants. The lesson from this is that it is just as important to measure the amount of 'benefit' a patient gets from an implant as to measure their 'performance' while using it.

What of the future? So far as our patients are concerned, they would like a fully implantable device with no external parts, an implanted microphone, possibly using the tympanic membrane itself as the head of a microphone, and with batteries rechargeable through the scalp. Such batteries would need to be removed for cremation.

At present a great part of each implant is made by hand. Electrode terminals, for example, are soldered to their wires under a microscope in an especially clean environment. Manufacture by machine would reduce the cost of making a device; at least one such technique for electrode manufacture already exists.

Apart from advances in implant design and speech processing, there are interactions between implants and other forms of new technology from which we can learn. The philosopher Ludwig Wittgenstein's seventh proposition reads 'Wovon man nicht sprechen kenn, Daruber Muss man schweigen'. I am sure you will agree, once you have got the gist of this, that it should be engraved on committee room tables in every hospital and medical school in the land, especially when you know that it means 'whereof one cannot speak, thereon one must remain silent'. I apologise if this turns out also to apply to surgeons trying to describe advanced technology, as I shall do next.

Dr A. L. Giraud is a scientist who spent some of the last year in our Cochlear Implant Programme and at the Imaging Department of the National Hospital for Neurology and Neurosurgery, Queen Square, in London. With Professor Frakowiak of that institution, she studied a group of our implanted patients using positron emission tomography (PET).[24] PET scanning is used to identify the increase in blood flow in specific areas of the brain while these areas are active. For example, the visual cortex of the brain shows an increase in blood flow while a subject is looking at a landscape. This change also occurs if, with your eyes shut, you imagine a familiar scene.

The centres or cortexes of the brain which deal with specific senses: hearing, touch, smell, vision, had traditionally been thought to keep themselves very much to their own specific sensory modality. Recent evidence, however, suggests that the situation is more complex. A subject who has become blind and learned to read using Braille continues to show an increase in activity in the visual cortex while using their sense of touch to 'read'. It is also well established that unemployed areas of sensory cortex can be utilized by modalities different from those normally represented in that cortex. For example, in animals brought up deprived of vision, parts of the unused visual cortex are taken over by the senses of touch and hearing.

Twelve adult patients in our Cochlear Implant Programme took part in a study using PET scanning. There were two groups of six patients. Six were studied during the first week after activation of their cochlear implant ('switch-on') when they had just begun to use their devices. A separate group of six adults consisted of experienced, successful implant users who had been using their implant for between one and three years. In addition, there was an equivalent group of six experienced French adult cochlear implant users, studied earlier at the Lyon Cochlear Implant Programme.

Figure 12 shows three 'virtual brains' all listening to words through their cochlear implants via a loudspeaker. The patients were not lip reading during any of the tests. The upper two 'brains' are drawn from a set of controls: volunteers with normal hearing. While these volunteers with normal hearing listened to words their visual cortices are not in a state of activity. The lower 'brain' represents the average of 12 experienced cochlear implant users. Their visual centres, shown on the left of the image (the brain is seen from the side, with anterior to the right and posterior to the left) show an increase in activity while the subjects listened to words.

Figure 13 shows the difference between naïve users, with only one week of implant use, and experienced ones, who had been using their implants for between one and three years. Dark red represents a weak signal, and pale yellow or white is a strong signal, again in the visual centres. The brain on the left of the figure represents naïve patients soon after activation of the device. There is a response from the visual cortex, but it is relatively weak. Moving to the right, in the next two brains, representing experienced cochlear implant users, the visual centre shows a significantly stronger level of activity.

Figure 14 shows a diagrammatic slice of the brain in the axial plane, in the lower of the two pictures. This shows the relative positions of the auditory cortex (labelled 2) and the visual centre (labelled 1). The auditory cortex has two parts. The primitive part simply 'hears' sound. The other 'interpretative' part recognizes and also understands words, sentences and familiar sounds.

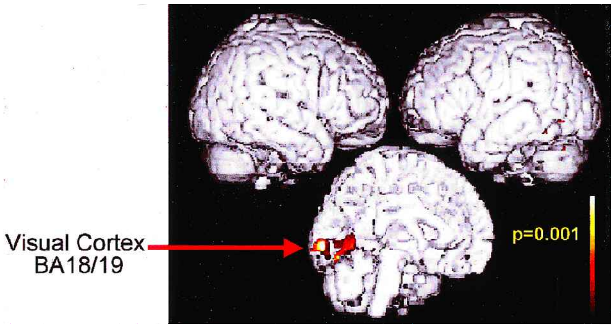


FIG. 12

(Colour plate) Upper two figures are from normal controls. No visual cortex activity is seen. Lower figure shows composite scan from 12 experienced cochlear implant users. Visual cortex activity is shown in colour. From: Giraud AL, Price, Graham JM, Truy E, Frakoviak RSJ. Cross-modal plasticity underpins recovery of human language comprehension after auditory reafferentation by cochlear implants. Reprinted from *Neuron* 2001;**30**:1–20, with permission from Elsevier.

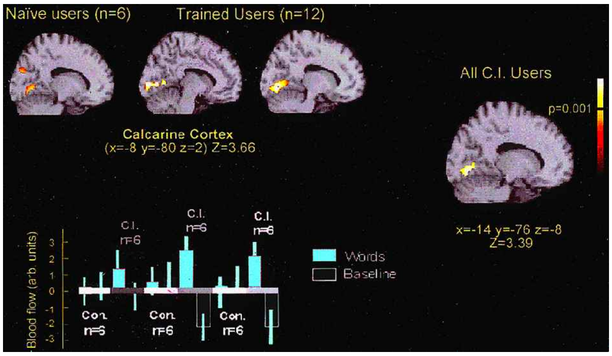


FIG. 13

Comparison between visual cortex activity in naïve cochlear implant users and experienced users. From: Giraud AL, Price, Graham JM, Truy E, Frakoviak RSJ. Cross-modal plasticity underpins recovery of human language comprehension after auditory reafferentation by cochlear implants. Reprinted from *Neuron* 2001;**30**:1–20; with permission from Elsevier.

The upper, right-hand graph represents the activity of the primitive auditory cortex. The blue blobs represent the response of this part of the auditory cortex to meaningless noise, while the red blobs are meaningful sounds and speech. During three years of cochlear implant use, moving from the left of the graph to the right, the signals all become stronger, represented by an upward shift on the graph, but there is no significant difference between the response to meaningless noise (blue) and to meaningful sounds and speech (red). The lower graph on the right is from the part of the auditory cortex that recognizes speech and familiar sounds, rather than simply 'hearing' them. There is the same progressive increase in the activation of this centre over three years, but also an increase in its ability to distinguish meaningless noise (blue), from speech and familiar sounds (red), which are shown significantly higher in the graph. This is probably what one would expect: with increasing practice, the brain seems to become better at interpreting auditory signals.

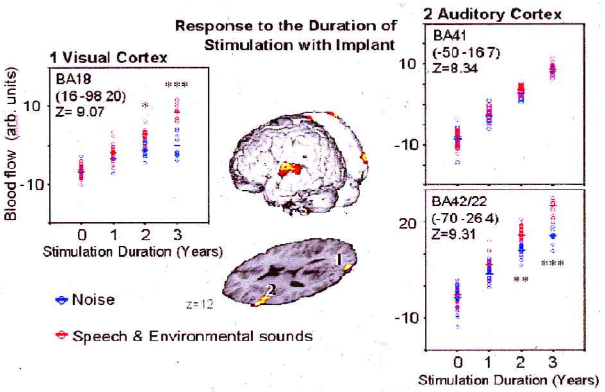


FIG. 14

Changes in visual and auditory cortex activity during the first three years of cochlear implant use. Discrimination between responses to meaningless noise (blue) and speech or meaningful environmental sounds (red). Measurements from the visual cortex are shown in the graph on the left; those from two parts of the auditory cortex in those on the right. The upper auditory cortex graph represents the 'primitive' auditory cortex, the lower graph represents the 'interpretative' auditory cortex. A parallel increase in discrimination between meaningful and meaningless sound occurs each year in the visual cortex and the 'interpretative' auditory cortex, but not in the 'primitive' auditory cortex. From: Giraud AL, Price, Graham JM, Truy E, Frakoviak RSJ. Cross-modal plasticity underpins recovery of human language comprehension after auditory reafferentation by cochlear implants. Reprinted from *Neuron* 2001;**30**:1–20, with permission from Elsevier.

Remembering that all the subjects were successful implant users and that the tests were performed without lipreading, it might have also been expected that as, with the help of their implant, they became accustomed to using auditory clues alone, without needing to lip-read, the role of the visual centres would fade. This turns out not to be the case. The graph on the left shows the activity in the visual centres: there is a change in activity comparable with the one that occurred in the 'recognition and understanding' part of the auditory cortex. There seems actually to be a progressive increase in the involvement of the visual centre, in parallel with an increase in the ability of the auditory cortex to differentiate between meaningless noise and familiar sounds.

Why should this happen? Djourno's patient found that when his cochlear nerve was electrically stimulated, all the frequencies above 1 kHz (about the middle of our range of hearing for speech) sounded alike. In English and French, consonants are represented by these higher frequency sounds. This means that Djourno's patient would have found it hard to distinguish between consonants because they would all have sounded alike. On the other hand, consonants are much easier to lip-read than vowels, which are represented deep in the larynx: if you block your ears and try to distinguish between a, e, i, o and u (and ou in French) using lip-reading alone, this task is more or less impossible. The PET results would therefore fit the hypothesis that the visual cortex of a deafened adult cochlear implant patient, with recent experience of lip-reading, nurses along its neighbour, the auditory cortex, in the first months and years after implantation, while the auditory cortex is trying to regain its old skills.

In this lecture I have tried to put the present state and recent past of cochlear implantation into historical context and trace a few historical links through the last five centuries. I have also tried to show why this field is so rewarding, fascinating and fun.

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