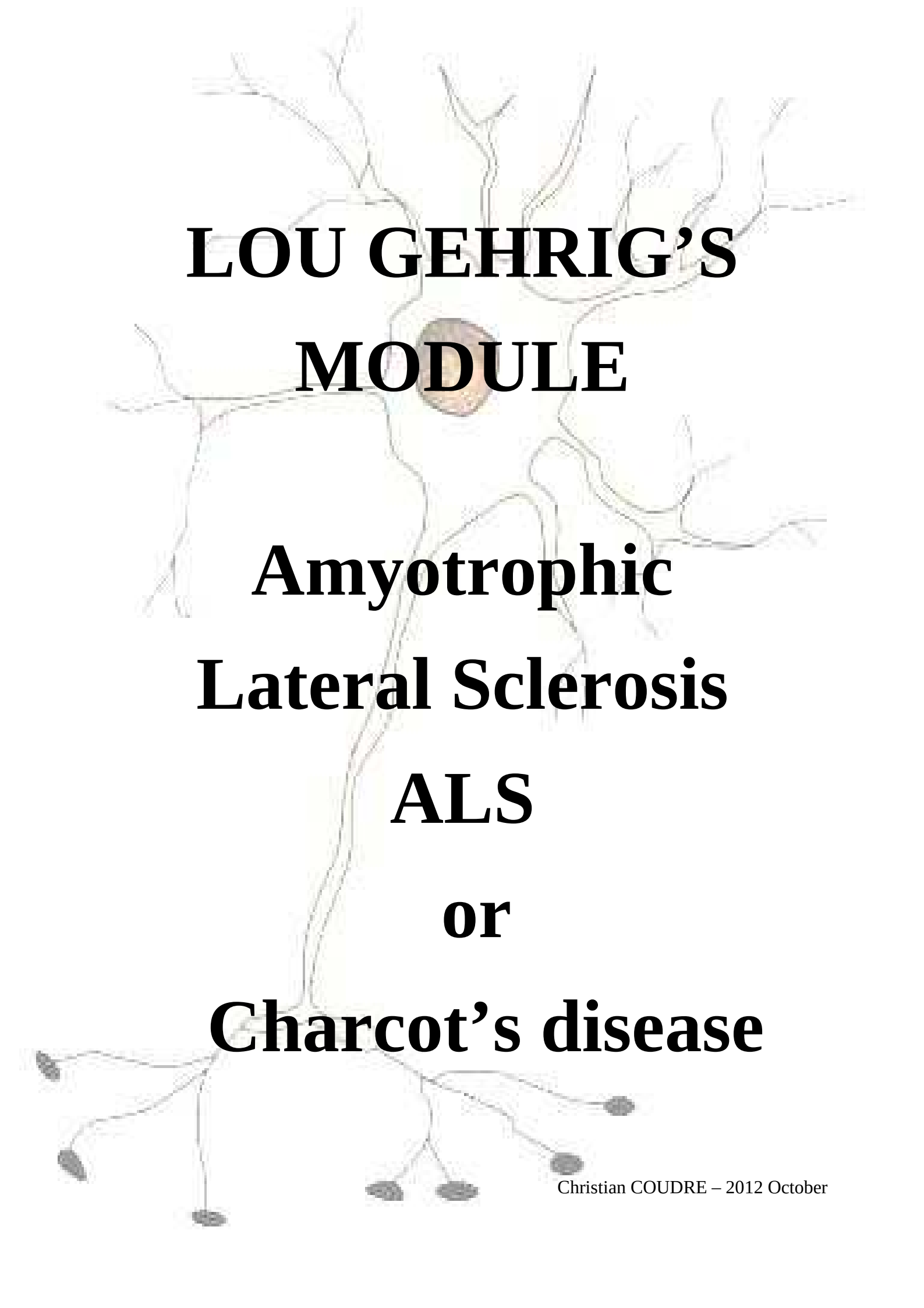

LOU GEHRIG'S MODULE

Amyotrophic Lateral Sclerosis

ALS

or

Charcot's disease

Christian COUDRE – 2012 October

Lou Gehrig’s module

Is ALS present in each medical and paramedical teaching? Today, the report is that numerous of generalist doctors, nursing and nurse's aides, ignore everything of this disease.

This module, without claiming to be a scientific manual, is a summary of bases necessary for understanding the Amyotrophic Lateral Sclerosis (ALS), also referred as Lou Gehrig’s disease or Charcot’s disease, not drafted as a theoretical course, but expressed by a real-life experience of seven years of fight against the muscular degeneration and the total loss of my vital functions.

45 old, (July, 2005), the first symptoms appeared. Very active in this period: TPM instructor, teacher in strategy of maintenance and marine biologist, for the main part, I saw my life stopping to leave place for a new adventure, a permanent fight against the progressive loss of my vital functions. In front of such a change, the natural law "adapt or die" takes its signification. Having crossed all the stages of the spinal and bulbar forms, I’m trying to explain at best the evolution of the disease such as I lived it.

Jean-Martin CHARCOT

(1825 – 1893)

French neurologist and professor of anatomical pathology, he’s known as "the founder of modern neurology" and is "associated with at least 15 medical eponyms", including Charcot-Marie-Tooth disease and amyotrophic lateral sclerosis (motor neuron disease). Charcot has been referred as "the father of French neurology and one of the world's pioneers of neurology.

Born in Paris, Jean Martin Charcot worked and taught at the famous Salpétrière Hospital for 33 years. His reputation as an instructor drew students from all over Europe. In 1882, he established a neurology clinic at Salpétrière, which was the first of its kind in Europe.

Charcot's studies between 1868 and 1881 were a landmark in the understanding of Parkinson's disease.

During 1882, he also rehabilitates the hypnosis in his works on the hysteric paralyses: lethargy, catalepsy, sleepwalking and amnesia. Sigmund Freud was one of his pupils. His son, Jean-Baptiste Charcot (1867-1936), doctor also, was an explorer and an author of oceanographic works in the Polar Regions.

Contents

What is ALS disease?

The SLA is linked to a degeneration of the motor neurons, involving a progressive loss of movements of the lambs (spinal affection), being able to go as far as involving a total paralysis. This degeneration can also affect the respiratory muscles and those of the gulp (bulbar affection), which may require the assistance of machines which substitute themselves for the vital functions.

The most frequent case begins with a spinal affection and with a gap of a few months in several years, follows itself a bulbar affection.

The sporadic ALS:

They are the most common forms, affecting 90 / 95 % of the cases of SLA. The age bracket affected by the sporadic forms is situated between 40 and 70 years old.

We distinguish two types of sporadic SLA:

- The spinal ALS count 80 % of the cases of sporadic forms. It begins with the affection of a limb, being able to affect a hand, reducing gradually its capacity to the prehensility between thumb - index, but it can also affect in the first one lower limb, altering the movements until a total paralysis.
- The bulbar ALS count 20 % of the cases of sporadic forms and affects the voice and the gulp.

The family form:

- Much rarer, this form is hereditary and considered as such when two cases are declared within a family. It concerns from 5 to 10 % persons affected by ALS. Persons affected by this form are frequently much younger, some being hardly more than 20 years old.

ALS mechanisms:

ALS is a progressive neurodegenerative disease affecting selectively nerve cells in the brain and the spinal cord. The loss of movements is the consequence of degeneration, meaning a cellular death of motor neurons, nerve cells which check the voluntary muscles.

This concerns two types of motor neurons which are situated at different levels from the nervous system: the "central" motor neurons and the "peripheral" motor neurons:

Central motor neurons are localized in the brain at the level of a region specialized in limbs movements called the driving cortex: they receive the order to execute a movement and transmit it in the brainstem and in the spinal cord.

The peripheral motor neurons are situated in the brainstem and the spinal cord, directly connected with the muscles to transmit them the order to make the movement.

Source: ALS Ile-de-France network – Written by Pierre-François Prada

An example of evolution

The evolution of the ALS does not follow a well-defined model. On the contrary, of a slow progress in the lightning form, a respiratory form which quickly pulls the patient in situation of respiratory distress syndrome than in the spinal form which concerns the movements of the limbs. It is one of the most frequent cases which I am going to describe you, my case for 7 years.

In July, 2005, everything began with an embarrassment in my right hand, with a weakness of the pinch thumb - index in the awakening. The practice of games of skill, writing, the do-it-yourself, became precarious across weeks and months, while the muscular atrophy gained the right hand, the arm, then the left hand side and, very gradually, the legs. Walking becoming difficult, uncertain, after one year and some bad falls, I had to accept the end of my autonomy and the assistance of a person by my side. At the same time, I had to give up my vehicle and make me transport to my company by reducing it gradually the number of working days.

In December, 2006, the muscle structure of my lower limbs became too weak. The appeal to an electric armchair was imperative for my outside walks, and indoor with a smaller model. The approach and the seated position to work on my office were relatively comfortable. The small armchair (Gochair), was linked perfectly, slightly obliquely, in front of the computer which I used by the movements of the head, my right hand which cannot any more manipulate and use the click of the

mouse. This Gochair also allowed me to put myself in table to have a meal with my family and to be nourishing with a spoon as a baby. But how much it was important to be still able to share these moments of my intimacy! The bulbar form, succeeding or adding up to the spinal form, made harder and harder the absorption of food, mixed, then liquefied, quite as the aspiration of liquids with a pipe which needs for a muscular effort, which a valid person does not suspect.

At the end of 2008, I get ready for a permanent confinement to bed, with the utilization of a patient lift to go still sometime on the armchair of the lounge, then to make only some coming go to toilet

Year 2009 was the descent towards the total paralysis. In November, my nutrition having become insufficient, at the risk of causing an important loss of weight and a deficiency in proteins, vitamins… The appeal to the gastrostomy tube was inevitable, a vital choice to continue to fight against the disease. The pose of the button comes true in two stages: a temporary probe which I kept approximately two months, before the installation of the button (Mickey), after healing of the stoma (surgical deviation of a natural pipe).

Two months later, the atrophy of my respiratory muscles did not allow me any more to expectorate correctly the bronchial secretions and, inevitably, the respiratory accident had to arise. On January 17th, 2010, one Sunday evening, I made a respiratory blocking. During 3 to 4 minutes, very serene, Valérie held my hand then, a few seconds before, losing consciousness, I accepted the intervention of fire brigades. I woke up three days later in the emergencies of Martigues with a tracheotomy.

My ALS vision

The diagnosis of the SLA needs series of examinations. The electromyography (EMG) is the main investigation allowed to confirm an infringement of the motor neurons of the internal horn of the spinal cord to estimate the importance and the area affected. In the most current form, which I've been living for 7 years, everything begins with the report of a loss of movements. The prehensility becomes harder and harder and, until falls the diagnosis, the hope to remedy this "embarrassment", was intact. Between the result of the first EMG and the interpretation of the neurologist, passes by a lapse of time which, in front of the importance of the result, we could not wait anymore for opening the report of the examination and discovering on Web what waits for us, the cruelty of the reality: "life expectancy, 3 - 5 years". There, bewildered, we take ourselves a wall in full face!

The evolution is very variable. Certain patients walk, speak, breathe or eat with more or less autonomy during years, but we also speak about lightning form, where the loss of the vital functions arises in some month. The progressive loss of its job, its activities of leisure and simple things of the life we never really appreciated: to walk, to breathe, to drink, to eat… disappear gradually, leaving us somewhere the time to adapt us by using mechanical systems: mixing food, being assisted for mobility, expression … But all is only temporary and our body becomes sluggish. Many people qualify it as "prison".

Numerous are the therapies which consist in "delaying" the degradation of the muscular functions: physiotherapy,

balneotherapy, sessions of phonology, etc. Knowing that only a passive gymnastic (manipulations, mobilizations) is recommended, any muscular effort is to be banned, be only increasing the stress and the degradation of the contractile fibers which constitute the muscle.

The impossibility to express himself, when arises a pain or an hard itch, without being able to intervene, requires a particular attention on behalf of, family, nursing, nurse's aides, because of the communication doesn’t pass more than by the glance which, by force of circumstance, becomes more and more meaning. In our case, lose the power of speech without being able to compensate with the slightest sign, if it is not by some mimes, requires the appeal to substitution technologies, to begin with panels of icons and letters, until the systems of eyes tracking allowing to use a computer by the movements and the blinking of eyes. Even if they are very expensive, their reason for being is vital so much it’s important to be able to call up or simply express a problem, a simple pain or a respiratory difficulty.

Every person occurring with a patient ALS has to adapt its gestures, its manipulations and the installation in an armchair or a bed. Because of our muscular losses, supports can ache, until cause bedsores in case of prolonged position. To lift a limb by squeezing it in the hand can ache. At the end, the most delicate action is the positioning of the head. When the muscles of the neck weaken, the head can fall in all directions, what causes a feeling of insecurity which can be compensated only with a total confidence in nursing aids.

Disease and psychology

The appeal to a psychologist is systematically recommended by the medical staff for the patient and family. Everyone reacts differently in front of the disease, from the total depression to the refusal of the armchair or the substitutions to the vital functions, to the acceptance by fighting the fate. This help is sometimes beneficial, using or not antidepressants. But it’s undeniable that love and friendship get an essential role in the mental balance of the patient. Nevertheless, the most important reason to want to hang on to the life is to keep an activity. The technology allows, even the most advanced steps of the disease, to use all the functions of a personal computer for the persons confined to bed, due to the systems of eye tracking and other simpler adaptations for those who do not very feel at ease with the computing. Some people write books, create blogs, Web sites to share their passion, speak about the disease...

« Feel useful is to remain alive »

Keep the spirit active, wake up in the morning by knowing that we have something to do, make a commitment for a cause …

Physiological consequences

Beyond the muscular weakness, the losses of the vital functions pull the patient towards vital choices. There are two new steps to be crossed. In front of a nutritional deficiency due to a degeneration of the muscles of the gulp and an insufficient feeding, the patient has to accept for a stomach tube to bring to the body nutriments which it needs. Another choice, much more delicate, is the one which arises when the vital capacities become insufficient and put the patient in situation of respiratory distress syndrome. The only solution to live longer is the implementation of a breathing assistance by ventilation with a mask, or by the insertion of a tracheotomy if it’s necessary.

The patient who refuses solutions knows that the end of his life is near. He can die, either by a nutritional deficiency, or by a respiratory blocking due to the impossibility to expectorate the bronchial secretions.

Once the body is assisted by machines for the main functions, the life expectancy doesn’t have limit anymore as far as the medical supervision is adapted without defect. It exists, specific trainings for every type of machine.

The main unwanted effects of this disease are: spasticity excess saliva, localized itches, tracheal infections and edemas, blisters, bedsores …

Excess saliva is mainly due to the bulbar affection which prevents the natural gulp of the saliva. When the production of

saliva exceeds one liter a day, it’s only possible to reduce quantity with diverse tested treatments and not without side effects with, for example, the injections of scopolamine. When an unwanted effect becomes unbearable, we are ready to try everything only to make it bearable.

Itches, when they are deep and active, localized in several places simultaneously, are a real torture for the patient who has no possibility to relieve himself. When an itch is localized we speak about favorable zone, which can persist several weeks, several months, but which, most of the time, changes regularly of zone. In the case of ALS, this problem is often due to contacts with pillows, sheets… during long times. It’s neither eczema, nettle rash, nor an allergenic reaction. Still one of the mysteries of the ALS, necessarily linked to the muscular cast iron or to the immobility, but there is no efficient treatment.

Bacterial infections often provides from hospitalizations by placing a cannula (tracheotomy) or a stomach pipe. For example: staphylococci or pseudomonas bacillus we never suppress or with great difficulty. It is necessary to analyze secretions in every monthly change of cannula and adapt the treatment to the antibiogram. Many inconveniences accompanying the infections are essentially: tiredness, gastric acidities and unpleasant taste in mouth, especially in the awakening.

To avoid the cutaneous hurts, it’s necessary to vary as often as possible the position of the body, the limbs and to be attentive during toilet to any departure of hurt, edemas or blisters (pocket of subcutaneous liquid), to prevent of any escalation.

Mechanic assistance

My vital functions are now insured by machines. My life depends only on their reliability to perform their main function, but also to indicate any dysfunction.

Machines and functions:

- Feeding: is done by a volumetric pump which regulates the flow. This one must be adapted to the balance of the transit, the values being situated on average between 200 and 300 ml / hour. The food appears under the shape of a dough containing calories, proteins, vitamins necessary for the body and it’s directly injected in the stomach by the stomach tube (gastrostomy).
- The respirator controls inspirations and expirations. The configuration is realized by a specialist: duration of cycle, volume and must be perfectly adapted for the comfort of the patient. It’s possible that the device goes wrong further to a prolonged tracheal congestion or for any other dysfunctions. The respiratory rate can accelerate and become uncontrollable. It’s indispensable to have always two available machines in case of breakdown.
- The humidifier, also named "waterfall" has to inject in the respirator’s pipe an adjustable quantity of purified water. It’s not necessary to use the heater function which can cause an inconvenience. Nurse’s aide has to look constantly at the water level.

- The cough assist is used when the tracheal congestions are important and when a simple tracheal vacuum is not efficient enough. The machine feigns a cough and expectorates secretions.
- The oral vacuum cleaner or tracheal vacuum cleaner is a machine which inhales the saliva or the whites of the mouth and, for patients with tracheotomy, the bronchial secretions which appear with the bulbar form and increase with the evolution of the disease. When secretions are too thick or sticky, the inhalation can be facilitated by preceding it by a cough assist.
- The manual insufflator (balloon), always has to be nearby in case of breakdown respirators (hey yes, that possible!), or a big prolonged break of power supply. This device is very easy to use and never breakdown. Press simply the balloon in your respiratory rate, or every 4 seconds (16/17 cycles per minutes), then adjust.
- The generator is the ultimate security against prolonged power cuts. Even if machines have an autonomy of at least 3 hours with an internal battery. There is also a power supplier service which places first and foremost of intervention the persons dependent on assistance by electric devices (in France, but in others countries?).

Disease knowledge

For the nursing staff, nurses, generalist doctors, the SLA, as many other rare diseases, is only a swindle definition until the day when a real confrontation forces learning. They have to participate in specific trainings more adapted by professionals healthcare witch organize sessions in the use of certain devices as the cough assist and acts more invasive as the tracheal inhalation. It’s also desirable that the family is able to intervene in emergency situation, because the appeal to an outside help doesn’t still allow an intervention just in time

Tracheotomy

The tracheotomy requires particular care and nursing acts such as the replacement of the inner cannula or, more delicate, the cannula. Having a tracheotomy since January, 2010, I saw various approaches of the change of cannula. At first, it’s necessary to know that there are various types of cannula. But in the case of the SLA, the total loss of speaking and the respiratory autonomy leave only the choice of a cannula without window and with or without inner cannula. The replacement of the cannula is made once a month and the advantage to have an inner cannula is that by replacing it daily, the risk of fouling or to create an infection with germs is very limited.

The replacement of the inner cannula is a particular nursing act realized during the daily cares. By precaution, it’s advised to disinfect the cannula of substitution with Dakin and to take usual precautions, without exaggerating either with a ridiculous equipment of operating rooms! (Refer to medical manuals and courses in relation to this practice).

The replacement of the cannula can be realized in a hospital environment, in operating rooms or in simple rooms, but also in the residence for the patients “home care”. The minimum necessity is to be equipped with oxygen supply, a manual insufflator, sterile sets, and by precaution, in case of difficulties to insert it, a cannula of a lower diameter. Quite as for the replacement of the internal shirt, these precautions, without exaggerating either, aim at avoiding any infection and the introduction of new germs.

The practice of a tracheotomy and the installation of a permanent cannula with balloon cuff, deprive definitively the patient of the power to speech and the absorption of food, even mixed or liquid. It’s a very hard decision the patient mostly refuse at the first approach. Personally, I toke the decision few seconds before losing consciousness further to a respiratory blocking. I woke up three days later to begin a new stage. However, my words were already inaudible and I was, for two months, fed by a stomach tube. The adaptation is not boring and security brought by the respiratory insurance extends considerably the life expectancy.

L'Association

SLA aide et soutien

Having taken the measure of the isolation and the distress of numerous persons affected by the SLA on a social network, I naturally take the decision to create an association to help them and contribute to make known this pathology.

Identity card for SLA aide and support
on November 1st, 2012

Date of birth: April 18, 2012

SIRET: 751 470 808 00013

Number of volunteers: 22 / Steering board: 4

Number of memberships: 255

Programs of our association

- Bring a moral support and to fight distress and the isolation.
- Organize or participate in charitable actions mediatized for the benefit of research.
- Bring material aids to the persons affected by the ALS: equipment for communication, assistance for mobility or daily toilet and nursery.
- Publish this manuscript about ALS disease based on my experience, intended for the medical and paramedical education.