The Guillain-Barré Syndrome, A Personal Experience

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The Guillain-Barré syndrome (“French polio,” Landry-Guillain-Barré-Strohl syndrome, postinfectious polyneuropathy, or idiopathic polyradiculoneuropathy) involves spinal roots, peripheral nerves, and sometimes cranial nerves. The syndrome was described in 1916 by Guillain, Barré and Strohl, as an acute radiculoneuritis with acellular hyperalbuminosis of the cerebrospinal fluid. It was thought to be of toxic or infectious origin. The disease entity has been linked to a multitude of causes such as the Coxsackie virus, measles, Mycoplasma pneumoniae, infectious mononucleosis, the Epstein-Barr virus and the ECHO virus. The syndrome has also been reported to follow immunotherapy, a variety of vaccinations, and prophylactic inoculations. More recently, strong support has been found for the theory that the Guillain-Barré syndrome is analogous to experimental allergic neuritis (EAN). A lymphocyte-mediated hypersensitivity is etiologically important. The syndrome, however, has been known to develop during immunosuppressive therapy for prevention of rejection of transplanted kidneys, and in patients who have Hodgkin’s disease.

The pathologic manifestations of the Guillain-Barré syndrome are those of a demyelinating neuropathy with focal perivascular lymphocytic infiltrates. These lymphocytic infiltrates are found not only in peripheral nerves and nerve roots, but also within the central nervous system, and in the liver, kidneys and lungs.

The incidence is reported to range from 1.4 to 1.9 per 100,000, without any clear seasonal pattern or clustering. There does not appear to be any significant differentiation by sex, and all age groups can be affected. Mortality figures range from 10 to 27 per cent.

In more than half the reported cases, a history of a preceding upper respiratory tract infection is obtained, and infection with a virus or Mycoplasma may be demonstrable. The clinical features show wide variation; in the typical case, however, the onset of polyneuritic symptoms is rapid, with progressive but not always symmetrical weakness, usually starting in the legs and then affecting the upper extremities and the face. Bulbar and respiratory muscles are frequently affected. Because of the lower motor neuron involvement, the paralysis is flaccid, and the corresponding tendon reflexes are diminished or lost. The trunk muscles are less frequently involved, but in severe cases total quadriplegia may occur.

The muscles of inspiration are frequently involved, with resultant restrictive ventilatory insufficiency, necessitating mechanical ventilation. Of the cranial nerves, the facial is most often affected, followed by the sixth, third, twelfth, and fifth, in order of frequency.

Sensory disturbances occur mainly in the form of paresthesias, which are most marked in the distal parts of the extremities and usually precede paralysis. Pain exists quite often, in the form of headache, back or muscle pain. The muscles are tender to pressure, and deep sensitivity is affected; impaired position sense and loss of vibratory perception may occur. Characteristically, the various neurologic deficits are not parallel in severity.

A diagnostic feature of Guillain-Barré syndrome is increased protein in the cerebrospinal fluid without an abnormally high cell count. This “albumino-cytologic dissociation” is always present in Guillain-Barré syndrome. There is, however, no correlation between the increase in protein and the clinical manifestations.

Autonomic dysfunction occurs either as excessive or inadequate activity of the sympathetic and/or parasympathetic systems. Since the effective management of ventilatory insufficiency has become commonplace, the autonomic dysfunction has unquestionably contributed to the still high mortality rate in severe cases of this disease.

The treatment of Guillain-Barré syndrome is mainly symptomatic. The major hazards are respiratory insufficiency, autonomic dysfunction, and thromboembolic accidents. The value of corticosteroids in the treatment of this illness is not documented.

Usually the physician’s role with respect to human illness is that of an onlooker. As a physician who, due to Guillain-Barré syndrome, was quadriplegic, who underwent a tracheostomy and intensive physiotherapy, and who was totally dependent upon the ICU team, I would like to describe the hospital experience from the patient’s perspective. Initially, it was interesting to be exposed to the varieties of professional role-playing that patients sometimes
see. One's colleagues and friends become quite different characters when seen from the low angle of the horizontal position. The nurses assume a classification that does not appear in any nursing outline. In the eyes of the paralyzed patient, the physiotherapist gains a prominent position (a position that is probably far more elevated than he can imagine), when he patiently teaches him to walk again. Other personnel and technicians are divided into positive and negative categories. To explain these statements, let me describe events as they unfolded for me.

In January 1972, I was hospitalized for a week with a viral pneumonia, for which I received ampicillin, respiratory therapy, and physiotherapy. On the following Monday, after a three-day weekend of convalescence, I resumed normal working activities. Several phenomena occurred, but were attributed to the preceding viral infection. The hands and feet felt extraordinarily cold, despite the usual temperature, which was around 20 F. The feet felt not only cold, but also somewhat numb, especially on the plantar surface. Small discrete areas of paresthesia on the lateral aspects of both thumbs, near the nails, were noticed four to five days after discharge from the hospital. Paresthesias were more marked in the right hand, and were associated with slight tingling on the palmar aspects of both thumbs and all fingers; this sensation remained most prominent on the grip surfaces of the thumbs. With each succeeding day of the week the operating room corridors appeared to become longer and longer, and almost painful fatigue was apparent on Wednesday. By Thursday, it was practically impossible to get out of bed. The tingling in the hands got worse and became associated with a severe burning pain, first in the interscapular area and then spreading over the entire back down to the sacrum. These myalgias made it impossible to lie in bed on the back, and the only comfortable form of rest was to kneel in front of the bed with the chest supported on the bed. The burning back pain was soothed only by the application of cold, wet towels. On Saturday, the short walk to the bathroom could barely be accomplished, and the weakness of the leg muscles progressed into a total inability to walk. Muscle pain and pressure were felt when sitting on a chair or the toilet.

The diagnosis of Guillain-Barré syndrome came to mind immediately, and I was readmitted to the hospital exactly seven days after discharge following the viral pneumonia. Admission to the medical intensive care unit was thought to be necessary, because of rapidly increasing ventilatory insufficiency and decreasing ability to cough effectively. A tracheostomy was considered; vital capacity decreased rapidly.

It was rather interesting to observe the attitudes of the various medical specialists toward a colleague with increasingly severe restrictive ventilatory insufficiency. One suggested the use of a rocking bed, another the "iron lung," and yet another tried to make the prospect of a naso-endotracheal tube appear attractive. Finally, one requested an effective cough, and since this could not be accomplished by the patient, a tracheostomy was inescapably indicated, and the above-mentioned alternate procedures were abandoned. The tracheostomy was accomplished under general anesthesia without complication, despite a deviated trachea due to thickened pleura of the entire left lung. The ensuing management of ventilation consisted of short-term ventilator support, followed by spontaneous ventilation with humidified oxygen-enriched air.

The tracheostomy fostered the division of the nursing personnel into two groups: Those who could carry out tracheobronchial suction effectively without causing unnecessary and painful movement of the tracheostomy tube, and those who made aspirations an excruciating experience.

The psychological despondency due to paralysis and being unable to maneuver by oneself, compounded by the inability to speak or communicate, was relieved only by the fact that two ICU nurses were rather accomplished lip-readers. Essentially, the nurse taking care of a patient in this totally dependent condition becomes the most important figure in this extremely limited and frustrating life situation. The world has become small; there is a limited visual area in and from the ICU cubicle; one's own pains and physical problems, of course, cause overwhelming concern. Thoughts of the future may not be encouraging. Under these circumstances, one divides all of the people with whom one has contact into positive and negative groups. Despite the success of one's attempted detachment, the nurse who fulfills his or her duties effectively, and who is, in addition, empathetic, is far more welcomed. Her shift is gratefully anticipated, in contrast to that of the efficient but sometimes indifferent nurse who merely fulfills an obligation.

It was rather interesting to observe confreres from the various medical disciplines in their activities and attitudes. No matter how detached one tries to be, a vital capacity of 600–700 ml and continued excruciating, burning pain in the back and in all other pressure areas tends to cause one to form likes and dislikes quickly. Each of the consulting physicians,
experts in their fields and frequently good personal friends, did contribute to the fragmentation of care. They each made their visits, reviewed, examined, and contributed. Often, though, a schism would arise among the various disciplines and specialists. Rarely can the various consultants in general and internal medicine, chest medicine, neurology, otolaryngology, and physical medicine interact and truly form a team. Occasionally, they actually seemed to be in competition. This may be very well and good from an academic viewpoint, but because of this fragmentation the patient may suffer. Often it is the coordinating senior resident who becomes the catalyst of all good measures, and the nurses assigned to the patient who are able to make life bearable. Other important services are provided by the physiotherapist and respiratory therapist, and their continuous service and observation frequently provide needed encouragement.

As my illness progressed, the appearance of a left facial palsy made swallowing and the management of saliva somewhat uncertain. This, in addition to the relentless burning pain in all pressure areas, and a peculiar sensation of lying on wrinkled sheets or on a lumpy mattress, paralyzed, tracheostomized, and with an indwelling Foley catheter, made life, to put it mildly, somewhat narrow. The hands felt hot, prickly and burning on the palmar aspects, and the sensation was as if they were stuck into hot, moist, sharp sand. The only relief was obtained by putting the palmar surfaces on the cool metal rails of the bed. The feet felt as if they were in a bucket with hard ice chips. Socks and warming devices had very little effect, and the pressure pain was not relieved by lamb skins and frequent changes in position.

The temporary lack of bowel control became a physical and psychological dilemma, as did the severe constipation, which necessitated frequent enemas. In this situation the important issues became: Who is connecting the tracheostomy tube to the humidifier hose? When am I going to be turned, and by whom? Who will provide the bed bath in the morning? Is a back rub going to be provided to grant short-term relief from the burning pain? These were the important questions of the moment, and all other considerations became pale and insignificant by comparison. For the patient in this sort of condition, day and night become one, and sedatives, antihypertensives, and analgesics cloud one's consciousness. Noises and voices are interpreted anxiously, and one is overwhelmed with gratitude when the "right" nurse walks around the corner.

The psychological effects of the critical care milieu on patients have received a fair amount of attention. The presence of all types of monitoring equipment, the many people providing care, and the limited contact with the outside world, form a unique environment. One fails to understand why different blood samples have to be taken at different times and why the collection of the specimens is not coordinated. It seems incomprehensible that the x-ray technician always comes just when one is almost asleep, and then frequently has to repeat that chest film with a cold cassette, and with seemingly limited concern for the patient. One begins to wonder why the visiting consultant, who is supposedly a good friend, leans against the foot of the bed and bounces against it, causing rather severe discomfort. The annoyance with housekeeping personnel is blown quite out of proportion when the vacuum cleaner smashes against the foot of the bed. The nurse who whips the curtain aside makes the cracking noise of a shot, and the name plate stamper operated by hand in the middle of the night sounds like a rock-crushing mechanism. Inexplicable as these annoyances and disturbances may be at the time, they are far overshadowed by pain, curiosity, or fear when, after having been turned, one hears from the EKG monitor a prolonged tachycardia or other dysrhythmias.

Nevertheless, these rapidly changing impressions and influences, comforts and discomforts, vexations and consolations, become minor compared with the psychological reactions following discharge from the intensive care unit to a regular ward. An overpowering feeling of complete letdown was, in my case, associated with the return to the ward after removal of the tracheostomy tube. An almost paranoid feeling of being deliberately neglected overcomes one, and even the nurses' aide who brings ice water at 2:00 A.M. is a welcomed visitor. When a nurse from the ICU visits, she appears to be an angel from another world.

The daily physiotherapy in the swimming pool became a truly refreshing and uplifting interruption of the daily routine. I will never forget the gratitude I felt toward the physiotherapist when I was, still paralyzed, lowered into the pool on a stretcher top. The buoyancy of the warm water allowed me to walk gingerly between bars. The daily improvement of muscle function in physiotherapy was elating, and the efforts of the physiotherapist were greatly appreciated. However, despite improved muscle function, pressure on muscles such as on the masseters, the large thigh muscles, or muscles of the arm (blood pressure cuff) was still painful and, in fact, is still bothersome four years after the onset of the illness.

I have discussed muscle pain and burning sensations. These were associated with other peculiar
sensory manifestations such as numbness of the skin over the entire chest, most marked over the left hemithorax. Light touch was felt as through a firmly clinging shirt, and a feeling of a restriction of intercostal and chest muscles existed, being particularly noticeable upon breathing deeply. It seemed as though the previously very painful muscles, be it those of the trunk, the back or the intercostal muscles, now resisted the stretch of deep breathing. The entire left chest felt as if it were covered with adhesive tape. A different sensation existed over the abdomen, where at one time early in the disease no sensory stimuli were perceived. Later, this was followed by hyperesthesia from T10 caudad, and during recovery this area too felt as if covered by adhesive tape. The muscles of the face, particularly on the left side, produced some discomfort when stretched upon yawning or eating. In the recovery phase, when muscle strength was regained, eating was almost always associated with an increased secretion of nasal fluid, which was much more marked from the left nostril. This phenomenon persists to this day, but seems to be slowly decreasing in intensity.

The physician–patient is frequently considered difficult to manage because of his knowledge, partial as it may be, and his professional standing. I made every effort to be "a good patient," not inquiring unduly (which was not extremely difficult with a tracheostomy), not questioning any measures, and abiding by all the rules. This "modus operandi" was adopted, not only because of my innate good nature, but also because previous patient experience had taught me that, in general, a positive and complying attitude is the most beneficial for everyone concerned.

To some extent I felt embarrassed to cause decision-making problems for my friends and colleagues. It is so much easier to be objective and detached about a stranger; when the patient is a friend, emotional involvement may, in some instances, cause mollification and delay of decisions. I have no criticism for the physician–friends who treated me, and no quarrel about the care I received from the entire team. The few disturbing facts that I have described are brought out merely to point to some weak links in the basically strong chain of their efforts, and to indicate possible areas of improvement. It was at their behest that I undertook to write down some of my reactions and responses to my hospital experiences. It is with a sense of profound gratitude that I complied with their request for the benefit of our patients.