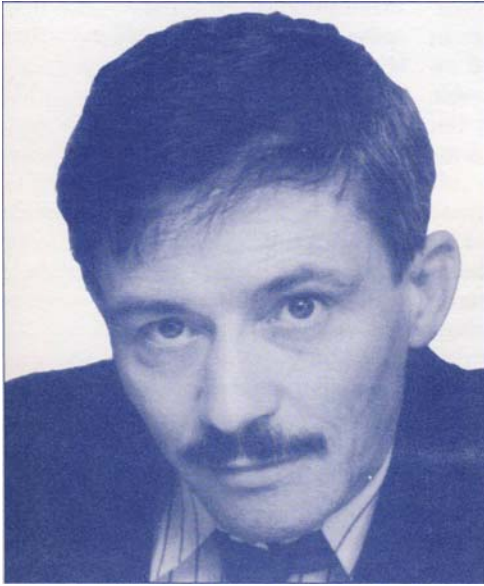


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They told him the disease was fatal; that no one knew its cause or treatment. But Claude was too young and spunky to say die. He decided to do something about it — not just for himself but for others like him.

PHOTO: GILLES LAFRANCE

# Claude Saint-Jean's Race Against Time

BY FRANCINE DUFRESNE

THE first faint light of day greeted Claude Saint-Jean as he walked across Montreal's Concordia Bridge to his summer job on St. Helen's Island where Expo 67 was in full swing. In a few hours, the bridge would be swarming with people; now, shortly after dawn on June 15, 1967, it was deserted. No one was there to see the 15-year-old youth suddenly lose his balance, sway back and forth, then halt as if he had been nailed to the wooden planks.

Claude felt no pain. He could hear and see clearly. But he couldn't control his limbs. They moved or they stood still, regardless of what his brain commanded. He seemed to be living a nightmare in which he was frantic to flee, but could only stand helplessly rooted to the spot.

Then it was over. Claude was in full control again and continued on his way, not sure what the trouble had been. In fact, he had just felt the first symptoms of an insidious and fatal disease.

Claude felt fine ail that day at the restaurant where he worked as waiter, cashier and kitchen helper. But the symptoms reappeared the next day, the day after that, every day, two or three times a day. Sometimes the attacks came at work. Luckily they were always so mercifully short — only a matter of seconds — that nobody seemed to notice.

At home, Claude said nothing at first. His parents were already worried about his older sister Francine, 17. Five years earlier she had been stricken with an illness called Friedreich's ataxia and was now confined to a wheelchair. Little was known about the disease except that

it affected the central nervous System of apparently healthy victims and was accompanied by degeneration of the spinal cord. But Francine, Claude reassured himself, used to fall down frequently when her illness began. He had no such trouble.

After two weeks the attacks became more than he could suffer in silence, and he confided in his mother. She sent him at once to a doctor, who attributed the symptoms to overwork.

Not satisfied, Claude went to a neurosurgeon. A week later, in Maisonneuve Hospital, a geneticist pronounced the dreaded verdict: Friedreich's ataxia. The treatment? Claude was advised to rest — and be patient.

But the boy refused to accept such a passive attitude. With his father, Claude went to another doctor, an old family friend who, although not a neurologist, had undertaken to find out ail that medical science knew about Friedreich's ataxia.

Calmly, gently, the doctor gave him the terrible prognosis. Friedreich's ataxia, a hereditary disease, is slow to develop, but irreversible. Besides losing his balance, Claude would become clumsy, and his hands would tremble uncontrollably. His speech would become impaired, sometimes unintelligible. Eventually, he would be confined to a wheelchair, then to a bed, unable to move or speak. Death would come finally from heart failure or infection. Claude probably had approximately a dozen years to live.

"Surely something can be done," the youth pleaded. "Something that could save me."

"I'm sorry," the doctor said. "There's no known treatment for this illness."

Now came devastating anger: Claude smashed his brother's 35-gallon aquarium into a thousand pieces. Then despair: Grabbing a kitchen knife, he barricaded himself in the bathroom. Alarmed, his distraught parents called the police who managed to calm the hysterical youth. He was then taken to St. Jean de Dieu Hospital, an institution for the mentally ill.

Claude remained in the observation centre for two days, just long enough to become friends with Dr. Marie Caron, a psychiatrist. She continued to see him and, bit by bit, helped him learn to live with his illness, encouraging him to keep up his studies and live as normally as possible.

Now with a grip on himself, Claude began to keep a precise diary of his symptoms, feelings and daily activities. But that wasn't all. Wanting to find out exactly how far research into his disease had gone, he visited every neurologist he could find. He even wrote to the federal and provincial health ministries. But always he got the same sad answer. There had been only two significant research studies of Friedreich's ataxia; one in England, the other in Sweden. Ail they produced were hypotheses. The cause of the disease remained a mystery.

Claude next decided to take matters into his own hands. He would spur research into Friedreich's ataxia. It might not do him much good, but if he could do anything to hasten the day when future sufferers might be cured, he would. That would be his goal from now on.

And so, passionately, persistently, Claude pleaded with one specialist after another to give some time, no matter how little, to the study of this little-known disease. Among 28 carefully chosen targets, he gave special attention to Dr. André Barbeau, director of the Neurobiology Department of the Clinical Research Institute of Montreal, internationally known for his part in the development of L-Dopa as a treatment for Parkinson's disease and for his work on epilepsy.

"Frankly," admits Dr. Barbeau today, "I quickly tired of the young man's repeated and time-consuming visits on a seemingly hopeless mission. I told Claude that it would be unrealistic of me to throw myself into costly research for such an uncommon disease."

Claude's reaction was simply to dig for more facts. He learned, for example, that Friedreich's ataxia was not, in fact, ail that uncommon. As of 1971 more than 250 persons were suffering from it in Quebec alone; what's more, the number of cases was on the rise. Claude could deduce this from the reports he pieced together from medical sources around the province and, tragically, he could see it within his own family; in 1970 his younger brother, Luc, started to feel the first symptoms of the disease.

Now Claude began to send letters to the United States and Europe in search of information. In May 1972 he presented Dr. Barbeau with a thick file crammed with data and medical opinions from two continents.

The famous specialist was not as impressed as Claude had hoped.

"Listen, we're still wasting time," Dr. Barbeau told him. "If you really want to go about this the right way, set up an organization and go after money for research."

Claude lost no time in beginning his campaign. In July he appealed for public support in a letter to Quebec's French daily, *La Presse*. By December the Association Canadienne de l'Ataxie de Friedreich, Inc., (ACAF) was officially chartered. With 250 members it became the first organization in the world to raise funds for research into the disease.

At its inaugural meeting on March 18, 1973, ACAF elected Claude Saint-Jean president, and his father, Leonard Saint-Jean, associate-secretary. By October 1973 Claude's relentless efforts had led to the formation of a scientific committee and team of 65 specialists – neurologists, cardiologists, biochemists, geneticists, paediatricians and psychiatrists – to carry out research into Friedreich's ataxia under the chairmanship of Dr. Barbeau.

It was the first body ever to unite researchers from Quebec's four medical schools – Laval, Sherbrooke, McGill and Montreal – in pursuit of a common goal. They decided first to establish just how prevalent the disease was in the province and then to examine the possibility of tracing all the cases among French Canadians back to common ancestors.

Because most of Quebec's five million French-speaking inhabitants are descended from 17,000 17th-century settlers and some of them have remained isolated geographically and largely homogenous (inter-marriage has been common), the committee looked upon Quebec as an ideal place to unravel the mystery of Friedreich's ataxia. They knew that the disease is transmitted only when both parents carry it. Although Mr. and Mrs. Saint-Jean have no known blood relationship, one victim out of every five is the child of parents who are related.

Today the committee is working with a group of 100 patients who constitute the most significant sampling of Friedreich's ataxia cases ever studied. Although the researchers donate their services, there are technical and administrative costs. In the last fiscal year, ACAF had only \$50,000. "It takes nerve to set up this kind of research with such a modest budget," says Dr. Barbeau. "But Claude perseveres like the very devil."

Dr. Barbeau is confident the action will get results. "A researcher," he says, "is a little like an artist who plods along until, all of a sudden, the moment comes when he is sure that his work is going to mean something." Barbeau claims he is beginning to feel just as he did in the months leading up to the discovery of L-Dopa therapy in 1961. His committee's work is opening up the possibility of treatment, he says, not only for Friedreich's ataxia but for other forms of ataxia — as well as diseases such as multiple sclerosis — that impair bodily coordination.

Claude Saint-Jean, meanwhile, wants to be a doctor and perhaps some day join that team of researchers. It is a race against time. It's been an effort for him to talk and to move ever since he entered medical school in the fall of 1973, and he lives in constant fear of a sudden deterioration in his condition.

Even so, his studies aren't enough to keep him busy. Claude and his brother Luc, a psychology student at Quebec University, have been setting up new branches of the association, writing briefs for other organizations interested in helping, and visiting and keeping in touch with victims of the disease throughout Canada. The team also puts out an information bulletin for ACAF members called *L'Eldorado*, a name meant to signify a better world, one full of hope and fabulous discoveries.

ACAF headquarters is the Saint-Jean family home in the Montreal suburb of St. Leonard. From his basement office, Claude directs its activities. Slender, handsome, elegant, he fixes his interviewer with his clear blue eyes. "Fm impatient," he says frankly. "Almost violently so. I'd like the research to be moving faster, the researchers to be doing more. Dr. Barbeau scolds me for this."

The Reverend André Reny, a Catholic priest who has known and admired the Saint-Jean family for eight years, comments: "Francine, Claude and Luc are all full of get-up-and-go

and, considering everything, very self-reliant. Claude, in particular, gets through a work load that few people blessed with full health could handle. He is a guy who can stir people up and make things happen. I'm not thinking only of his efforts to promote research. He's just as concerned about the other purpose of his association: to make contact with ail the victims of his disease, to let them know they're not alone."

Indeed, Claude spares nothing of himself to help those in the same boat. He gives frequent radio and television interviews, talks to students and service organizations and so far has written more than 10,000 letters by hand. "What's even more remarkable," says journalist Lizette Gervais, "is that Claude readily submits to medical examinations and tests of ail kinds, acting the part of guinea pig with endless patience."

And so, Claude Saint-Jean today continues his merciless struggle against time. "I feel like a dog whose chain is gradually being shortened," he wrote in his diary one day. "But I have never accepted my disease. Logic tells me I won't make it. I know it's accelerated in the last two years. But at least I'll have the satisfaction of having tried everything, of having helped others, of having fought to the end. And who knows? Perhaps of winning."

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