Letter to the Editor

To the Editor

Re: The Growing Pains of Spinal Surgery

R.J. Hurlbert, Can J. Neurol. Sci. 2004;31;136

Dr. Hurlbert has written a thoughtful and diplomatic editorial regarding my article on spinal education in neurosurgery. I would like to respond to a few of his points.

Dr. Hurlbert has asserted that because none of the Canadian program directors are dedicated spinal surgeons, their opinions regarding resident spinal competency are invalid. Program directors are appointed to organize and implement six years of neurosurgical training across all neurosurgical subspecialty fields. Towards this end they exert considerable time and resources. How does a non-spinal program director evaluate a resident's spinal competency? He asks the dedicated spinal surgeon in his group. The same way he does for every other area for which he is not an expert – he relies on the opinion and commentary of the appropriate expert colleague. The Royal College mandates that Canadian program directors work with a specifically appointed Resident Education Committee, and this committee meets at least quarterly. Through this committee resident evaluations are made. For example, a vascular neurosurgeon/program director relies on his Resident Education Committee to evaluate a resident's competency in spine management, neuro-oncology, functional disorders, peripheral nerve competency, etc.; indeed he is able to sign-off the final in-training evaluation on this basis. To dismiss the study on the assertion that non-spinal program directors are unable to assess spinal competency displays a lack of understanding about the process by which program director's evaluate their residents.

The statistics regarding membership ratios in various spinal organizations has relevance to a number of issues, but not towards the necessity of teaching spine surgery to neurosurgical residents. If one wanted to use numbers in this regard, the more relevant figures are that almost 100% of graduating neurosurgeons will engage in spinal surgery of some description, whereas only a small fraction of orthopedists would do likewise. Hence the onus on neurosurgical residencies to ensure spinal expertise to its graduates. To do so, program directors indicate their need to maintain 'spinal surgery under the umbrella of neurosurgery', as opposed to delegating it elsewhere. I trust none of the program directors are embarrassed by this need. While it is appropriate and politically correct to 'recognize the richness and quality of input'of both neurosurgeons and orthopedists, as suggested by Dr. Hurlbert, in terms of the day-to-day work of running a residency program, managing 'political parental umbrellas' is paramount. Parental umbrellas dictate clinical resources and thus clinical services and, like it or not, clinical services are the basis on which resident training is based.

I agree with Dr. Hurlbert's sentiment that it is unfortunate that there are no dedicated spine surgeons in the position of a Canadian neurosurgical program director. If there were, they too would understand the challenges of current resident education, and not just in spinal surgery. A program director is responsible for training the residents in all the subspecialties, while the subspecialties themselves seem to be distancing themselves from resident education.

The survey was not meant as an attack on the value of neurosurgical or orthopedic spine surgeons, which seems to be the defense of the editorial. Instead its theme was education and was meant to discern how neurosurgical program directors feel about the importance of spinal education, and how they are achieving their goals in this regard. As Dr. Hurlbert points out, spine is a major part of neurosurgical practice, and that is exactly why neurosurgeon educators must take ownership of that education, and ensure its quality and appropriateness.

Brian D. Toyota,
Vancouver, BC

To the Editor

Re: Variable Phenotype in Gerstmann-Sträussler-Scheinker Disease.

De Michele G, Pocchiari M, Petraroli R, et al.
Can J Neurol Sci 2003; 30: 233-236.

De Michele and colleagues[1] recently reported an Italian family with Gerstmann-Sträussler-Scheinker disease (GSS) suggesting that the variable phenotype does not appear related to the codon 129 polymorphism.

The most common mutation causing GSS disease is P102L-M129, which results in the substitution of proline to leucine in coupling with methionine at residue 129.[2] Mutation at P102L with valine at 129 (P102L-V129) has also been reported.[3]

Recently, we published another GSS Sicilian case,[4] with a P102L-V129 mutation of PRNP gene suggesting a high frequency of GSS in the island and a possible foundation effect for P102L mutation. Our patient showed psychiatric manifestations at onset with apathy and depression that are extremely rare in these patients; two years later, he showed cerebellar ataxia, psycho-organic syndrome, dysarthria and seizures; three years after the onset, he developed lower limb numbness and increasing gait difficulties, progressive weakness, swallowing and breathing difficulties and progressive cognitive decline with dementia. He died at 45 years of age, four years after the disease onset. His father and brother died with the same disease and the similar psychiatric onset (at 57 and 51 years of age respectively).

The age of onset of our index case suggests a familial genetic anticipation mechanism, not previously reported even if evident also in the published pedigree 1 (patients III-3 with IV-3 and III-12 with IV-11). The significance of this age anticipation appears, to our knowledge, evident but inexplicable.

The considerable clinico-pathological diversity of GSS is probably related, at least in part, to different PRNP gene mutations and discordance within families points to additional genetic and environmental disease modifying factors, including codon 129 status. Moreover the clinical course of our case[4] only

partially overlaps the reported phenotype associated with P102L-V129; therefore we think that a genotype-phenotype relationship may exist but it is difficult to explain on a simple genetic basis. We speculate that the polymorphisms may play a role in this relationship.

The inherited prion disease is a unique genetic model to illustrate how nonpathogenetic mutations can influence the phenotype caused by pathogenic gene mutations. This event could be consequent to altered protein conformation, even if other unknown additional factors might be considered as background for phenotypic variability.

Marco Bianca, Sebastiano Bianca, Carmela Ingegnosi, Ignazio Vecchio, Rocco Raffele, Liborio Rampello, Francesco Nicoletti
Catania, Italy

1. De Michele G, Pocchiari M, Petraroli R, et al. Variable phenotype in a P102L Gerstmann-Sträussler-Scheinker Italian family. Can J Neurol Sci 203; 30: 233-236.
2. Ghetti B, Piccardo P, Frangione B, et al. Prion protein amyloidosis. Brain Pathol 1996; 6: 127-145.
3. Young K, Clark HB, Piccardo P, et al. Gerstmann-Sträussler-Scheinker disease with the PRNP P102L mutation and valine at codon 129. Mol Brain Res 1997; 44: 147-150.
4. Bianca M, Bianca S, Vecchio I, et al. Gerstmann-Sträussler-Scheinker disease with P102L-V129 mutation: a case with psychiatric manifestations at onset. Ann Genet 2003; 46: 467-469.

Response

We read with interest the letter by Bianca et al, reporting the fourth Italian family with Gerstmann-Sträussler-Sheinker disease (GSS). Previous studies[1,2] and ours demonstrated variable clinical presentation, apparently not influenced by valine/methionine polymorphism on codon 129. In our family, patient III-8, with 'ataxic' phenotype, was homozygous for methionine on codon 129, and patient IV-11, with 'cognitive' phenotype, was heterozygous for valine/methionine on codon 129. Therefore, excluding the unlikely hypothesis of an intragenic recombination, we can infer that our patients had P102L-M129 mutation. The presence of the same mutation in patients with different phenotype is not in favor of a significant role of this polymorphism.

Onset age anticipation in GSS is an interesting issue, and it has been also reported in Creutzfeldt-Jakob disease linked to the E200K mutation.[3] It is possible that some genetic factors may influence the disease onset, however, we should point out that anticipation may be due to biological mechanisms, as for CAG triplet diseases, but may also depend on observational biases. Patients with early onset in older generations are less likely to be ascertained directely (because they are dead) or indirectely (because they do not reproduce), and patients with late onset in younger generations may have not yet shown the disease. Awareness of the disease may lead to earlier diagnosis in younger generations. To solve this issue we need studies which include a large number of families and which take in account all possible sources of bias.

Giuseppe De Michele, Giovanni Coppola, Alessandro Filla
Federico II University, Naples, Italy

1. Barbanti P, Fabbrini G, Salvatore M, et al. Polymorphism at codon 129 or codon 219 of PRNP and clinical heterogeneity in a previously unreported family with Gerstmann-Straussler-Scheinker disease (PrP-P102L mutation). Neurology 1996; 47: 734-741.
2. Tanaka Y, Minematsu K, Moriyasu H, et al. A Japanese family with a variant of Gerstmann-Straussler-Scheinker disease. J Neurol Neurosurg Psychiatry 1997; 62: 454-457.
3. Rosenmann H, Kahana E, Korczyn AD, et al. Preliminary evidence for anticipation in genetic E200K Creutzfeldt-Jakob disease. Neurology 1999; 53: 1328-1329.

To the Editor

It's not a looming crisis – the crisis is here, now!

Re: Manpower in the Canadian Neurosurgcial Workforce: Is a Crisis Looming?

J. Max Findlay, Can. J. Neurol. Sci. 2004; 31;138

We found it interesting to read the article written by Dr. Findlay in the previous issue of CJNS about the lack of opportunities in Canada for newly graduated neurosurgeons. He states that there is a 'looming crisis'– we feel that the crisis has already started. All three of us graduated in July 2003 and have been looking for jobs in Canada. Our job search has been unsuccessful so far and we are, therefore, considering opportunities in the USA.

Since 2000, the CCNS has warned program directors in neurosurgery that there will be an excess of neurosurgeons in Canada by 2004 and that new graduates will not be able to find employment. However, in the last four years nothing has been done to address this 'looming crisis'. In fact, the hiring of new residents in neurosurgery programs continues at the same rate as in the past. In addition, McMaster University was allowed to start a training program recently!

Why is nobody paying attention to this problem? We know that residents provide an essential and important service at a very negligible cost compared to certified specialists, making it very cost-effective for hospitals to continue hiring them. In addition, we think neurosurgeons working at tertiary centres with established training programs would find it very difficult to practice without residents. Residents take call, do all the 'scut' work, assist in the operating room, work-up and look after the medical problems of patients, and interact with both families and hospital staff to ensure quality patient management. In addition, a training program in neurosurgery gives the hospital and university a marketable 'status'.

We think that each training program is ethically bound to reassess the need for their program – and if they feel that it is necessary, they need to ensure that the graduating resident has a job. It would be immoral to accept and train neurosurgery residents knowing that they will not be able to work once they complete training in our public health care system.

It is only fair to warn future physicians contemplating neurosurgical careers that they should wisely consider the implications of enrolling in a neurosurgical residency program. In fact, current neurosurgery residents should probably reconsider their careers, and training in neurosurgery should be limited to a few select places in the country. Residents must understand that they may have to leave Canada to find work and that the USA is not currently an option. A solution for current

residents and medical students interested in neurosurgery is to complete their residency training in the USA to be eligible to write the American Board of Neurological Surgery examination and then come to Canada and write the Royal College examinations as US trained neurosurgeons. This would enable them to work in both the US and Canada.

There is little publicity given to the lack of career opportunities facing many medical and surgical subspecialists across Canada. In fact, even physicians are often unaware about the lack of jobs in certain specialties! It surprises them that some physicians and surgeons face unemployment once they complete their training if they opt to stay in Canada!

Ashok Modha, New York, NY, Richard Perrin, Toronto, ON
John Sinclair, Palo Alto, CA

To the Editor

Re: Manpower in the Canadian Neurosurgcial Workforce: Is a Crisis Looming?

J. Max Findlay, Can. J. Neurol. Sci. 2004; 31;138

Dr. Findlay has made astute observations regarding neurosurgical workforce issues on the horizon. His prediction of the unemployed neurosurgical graduate is quickly materializing as witnessed by those Canadian graduates still eligible for American employment, but who are either struggling to find any jobs in Canada in 2004 or are heading to the United States with extreme reluctance.

Accepting that the American Board of Neurological Surgeons' decision to bar Canadian training is fixed, one questions the obligation of Canadian neurosurgeons, and their 'governing' bodies, to be proactive about the looming crisis to which Dr. Findlay alludes. The only problem is that a governing body who has a responsibility to avert this crisis does not exist. Though many feel the neurosurgical specialty committee of the Royal College of Surgeons has this obligation, this is not the case. Rather theirs is one of setting standards of training and evaluating residency programs, among others.

Dr. Findlay predicts that the natural history for Canadian neurosurgical training will be a diminishing pool of medical school graduate applications, and hence each residency training program will contain a nominal number of house-staff. Most experienced program directors will admit that a minimum quorum of residents is needed to run the program optimally.

We should take Dr. Findlay's prediction one step further, with fewer applicants, what will happen to Canadian neurosurgical residency? Will Canadian neurosurgical training soon consist of many programs, each with a few residents? Or will it be a war of attrition, with only the bigger, resource-endowed academic centres that survive?

I suspect it will be the latter, and truthfully, from the perspective of the quality of training, I would suggest that this is the better path to pursue. In fact the Royal College neurosurgical specialty committee could indirectly orchestrate this movement by raising the standards of training. For instance, if the committee insisted that training in stereotactic radiosurgery, movement disorder procedures, epilepsy surgery, comprehensive peripheral nerve surgery and comprehensive spinal care are a minimum standard for program accreditation, many current Canadian programs would not exist. Perhaps this is a 'cut-throat' approach, but it has its advantages. I am doubtful that many academic Canadian programs would surrender their residency training as an altruistic move to avert the pending over-population of neurosurgeons. There is certainly no one with the authority to unilaterally direct a program to stop training neurosurgeons. Indeed, raising the bar in this fashion will enhance the quality of Canadian training beyond that of any other country – and if one accepts that the American Board of Neurological Surgeons decision was based on their unhappiness with Canadian neurosurgical education (and not job protectionism) – this may, ironically, be the avenue by which Canadian training is again accepted south of the border.

Brian D. Toyota,
Vancouver, BC

Response

Dr. Toyota makes several provocative and sensible suggestions in his letter, however impossible they are to imagine at the present time. Its seems unlikely that accredited neurosurgical training programs will voluntarily close, for all the reasons mentioned in the letter from Drs. Modha, Perrin and Sinclair. Despite reductions in their work hours and call duties, and even with the help provided by physician assistants or hospitalists in many centers, residents are still essential to academic neurosurgical units. We need them and, at least for now, we won't stop recruiting them, however uncertain their professional future is. I am not sure that this is "immoral", providing the trainees are properly informed, and the necessity of informing was really the main point of the editorial. As was also pointed out in my comment, there has been no shortage of candidates up until now.

Neurosurgical programs in this country have always relied on a steady exodus of neurosurgeons to the US. It has never been possible for this country to employ all of the neurosurgeons it produces. Looking back at my 14 years at the University of Alberta, nine of the 16 Canadian graduates of our program chose to practice in the US. The fact that that pasture is no longer much greener than our own (medical liability crisis there, the introduction of fair alternate funding or service remuneration plans here) and that graduates might, therefore, have recently become less inclined to move south, is irrelevant: residents are now ineligible to sit the American qualifying exams. Counting on work in the US as a "NonBoard certified" specialist is a very large gamble.

According to Dr. Modha et al, a manpower crisis in neurosurgery already exists and, as far as their experience is concerned, they are right. I am terribly worried about the future of all of our residents in training. We continue to lobby the American Board to reverse their decision, but since their ruling was in a large part based upon a perceived oversupply of neurosurgeons in the US (and Canadians stealing jobs from American trainees) it is difficult to fully articulate the cause of our protests. Many of us think that only a demonstrated shortage of neurosurgeons in the US will reverse their decision on this matter. Until then I, myself, will continue to discourage medical students from considering neurosurgery as a career, which is a terrible thing to have to do.

J. Max Findlay
Edmonton, Alberta