COGNITIVE PSYCHOLOGY OF NEUROLOGICAL ERRORS.
HOW SHOULD THE NEUROLOGIST GO-ABOUT MAKING A DIAGNOSIS.
THE MODUS OPERANDI

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Case example that will be used in my presentation.
RC. A 67 year old man. I was asked to see him in consultation in an Intensive Care Unit.

He had an aortic valve replacement several days before. On awakening he noted prickling and tingling in his hands and feet and severe weakness of his hands and weakness of his lower extremities. He does not feel this has changed since he first noted it when he awakened from the post-operative sedation.

He is alert and has no important abnormalities of his cranial nerves or cognitive functions. He has full visual fields. He has bilateral severe weakness of his upper extremities especially the hands- he has very little hand movement. His shoulders are weak but he can hold them up at shoulder level for about 5 seconds. His biceps and triceps are stronger than his shoulder girdle muscles and his hands.

He can lift his thighs off the bed but has considerable weakness of all lower extremity muscles including his feet and toes. His reflexes are brisk. His plantar responses are extensor. He has striking loss of position sense and vibration sense in his hands, wrist, knees, and maleoli.

He has diabetes Mellitus, atrial fibrillation, and chronic myelogenous leukemia. He was hospitalized after a motor vehicle accident 3 months before with some subarachnoid bleeding.

Diagnosis
The hunt for the diagnosis begins when the doctor greets the patient and observes and analyzes the patient's dress, comportment, mood, style, and body signals. Shaking hands and then watching the patient walk into the examining room yields further diagnostic clues. The effective Neurologist listens carefully as the patient describes his or her problems and self, and then formulates queries based on the initial history given and interprets the patient's responses to the physician's pointed queries, comments, and pauses. The Neurologist then conducts a meticulous general and neurological examination, testing clues, hunches, hypotheses, and theories generated by the history. Finally, the Neurologist uses information from laboratory, imaging, and physiologic investigations to arrive at a complete differential diagnosis and a quantitative estimate of the probability of various pathologies. The effective Neurological clinician is versatile enough to use any and all available information as clues to the diagnosis. In some patients, the diagnosis comes mostly from the history, in other patients from an unexpected finding on examination, and in still others from careful analysis of imaging and laboratory abnormalities.
The Neurologist must be systematic, penetrating, and thorough at making an accurate, medically sound diagnosis. The process of meticulous pursuit of the diagnosis, more than any other physician behavior, makes patients respect and trust their physician.

Methods and strategies of Diagnosis
Diagnoses are made mostly through three different but inter-related techniques: deductive reasoning, pattern matching, and inductive analysis. Deductive analysis involves sequential hypothesis-making and testing to deduce diagnoses. Deductive exploration should be both positive (looking for features and information that suggest or confirm a diagnosis), and negative (queries that might elicit features that argue against diagnoses). In my discussion I will discuss this approach because it is the one used routinely at the bedside or in the clinic during doctors encounters with individual patients. Pattern-matching involves comparing features of the history and/or the examination that match conditions that the clinician knows well, has seen, or has read about in the medical literature. Pattern-matching is largely intuitive.

Inductive analysis, in contrast, involves a thorough collection of data and then analyzing that data after it is collected for patterns and information that lead to the possibility of various diagnoses. This technique is retrospective and applies mostly to analysis of group data. As such I will not discuss this approach in this session.

First impressions gained by pattern-matching can be faulty. Doctors must beware of “anchoring in” and not questioning their initial impression of the pattern they think that they recognize. Many astute clinicians use a combination of cognitive strategies depending on the patient and the circumstances. Unfortunately many physicians do not pursue diagnoses in a systematic and thorough manner. Neurologists should become aware of and analyze their own diagnostic strategies and strive to improve them. One way to improve diagnostic acumen is to continue to review personal errors in diagnosis and analyze the types of errors made.

The Deductive approach to Neurological Diagnosis

1. The neurologist should always ask *what* is the disease mechanism and *where* is it located? These two queries should be pursued concurrently and from the very beginning of the patient encounter.
2. The effective clinical neurologist generates and tests hypotheses, gradually refining them at each phase of the clinical encounter. Hypothesis generation should begin early and should proceed throughout the encounter. During and after the history, during and after the general and neurological examination, and after each planned test or series of laboratory investigations, the skilled neurologist models and refines the anatomical and disease diagnoses much as a sculptor gradually chips away at the marble, slowly allowing a face to emerge.
3. The neurologist thinks in terms of probabilities, not absolutes. How likely is a particular diagnosis—80 percent, 50 percent? What are other possible diagnoses, and what is their probability of being correct? Also is there a diagnosis, although not a highly likely, that would be very crucial not to miss?

What data is used to answer the *What* question?

Disease mechanism refers to the pathologic entity or pathophysiology of the disease causing the patient’s symptoms, signs, and laboratory abnormalities. The major clues as to *what* is wrong come from the history. Data include:

The Demography of the Patient

Age, sex, and race affect the chances of a patient having a particular disease. A focal brain lesion in an elderly man has a different set of etiologic probabilities than a similar lesion in a young girl. Where the patient lives also affects probabilities. Multiple sclerosis is much more common in Minnesota and Ireland than in Northern Africa or Central America. Cysticercosis is more common in Mexico and Los Angeles than in Wisconsin.

The “Ecology” of the Patient’s Illness

A patient’s previous personal history and prior illnesses yield clues to the diagnosis. The presence of known lung cancer greatly increases the probability that neurological symptoms are due to direct spread or metastasis from the tumor or to an indirect effect of the cancer. An older man with a past history of coronary artery and peripheral vascular occlusive disease who develops a left hemiparesis has a high probability of having had a stroke due to either atherosclerotic disease of the right carotid artery or cardiogenic embolism. A patient with diagnosed systemic lupus erythematosus who becomes confused most likely has a steroid psychosis, a bleed into the brain caused by thrombocytopenia, or an infarct related to lupus anticoagulant or other coagulopathy.

The Family History, Occupation, and Exposures of the Patient to Various Risk Factors

These factors also affect disease probabilities.

The family history and genetic information can be of even greater importance after the history and examination have allowed the neurologist to localize the problem to a portion of the nervous system. The family history in patients with neuropathies, myopathies, and dementing illnesses can be the principal clue to a definitive diagnosis.

The Onset of the Present Symptoms

Some diseases begin abruptly during activity (for example, cerebral hemorrhages or embolism), while others are more likely to be noticed during the night or on arising (such as cerebral thrombosis or carpal tunnel syndrome).

The Course of the Illness

This is the often most useful item. A sudden onset brain lesion that is maximal at onset, improves, and clears quickly is most likely due to a brain embolus, whereas focal signs that develop and evolve during months and are gradually progressive are more likely due to tumor. Symptoms that progress gradually and inexorably during years are often due to degenerative disease. In multiple sclerosis, the neurologic deficit often advances during a period of days to a few weeks, stabilizes, then gradually remits, a time course unlike most other neurologic conditions. A course of illness graph can be a useful diagnostic tool.

Accompanying General or Neurologic Symptoms
Symptoms such as fever, jaundice, malaise, headache, seizures, and loss of consciousness also contribute heavily to accurate diagnosis.

What data is used to answer the *Where* question?
The most important information for localization of a lesion comes from the neurological examination and the results of imaging and physiological tests. The patient's account of the nature, as opposed to the timing, of the neurologic symptoms is also very helpful and may be the only available data in patients who have recovered from temporary symptoms. The patient may also recognize subtle deviations from normal when abnormalities are not evident on examination. Tingling, altered somatic or visual perceptions, and unaccustomed imprecision in speech are often described by patients even though the neurological examination is not sensitive enough to detect definite abnormalities in these spheres.

Some general localization principles:

Some Symptoms or Signs are Quite Specific for Certain Regions
Abnormal speech output characterized by the use of wrong words and poor comprehension and repetition of spoken language is diagnostic of a dominant hemisphere (usually left) temporal lobe lesion. Lights or unformed colored objects seen transiently but repeatedly in the left upper quadrant of vision with both eyes is diagnostic of a lesion in the lower bank of the right calcarine fissure.

The Distribution of the Particular Symptoms or Signs Helps Localize the Lesion
Weakness limited to the left abductor pollicis brevis, opponens pollicis, and first two lumbrical muscles is diagnostic of a left median nerve lesion. Tingling in the little finger and the medial half of the fourth finger that stops abruptly at the palmar crease nearly always indicates an ulnar neuropathy. Weakness of the left face and hand with increased left deep tendon reflexes and an extensor plantar reflex on the left means a lesion in or beneath the right precentral gyrus in the perisylvian area controlling the face and hand.

Combinations of Findings ("Fellow Travelers") Help Localize a Deficit
The strategy of trying to localize a lesion can be compared to trying to locate a car on a long road. Specific landmarks such as a restaurant, cross street, or traffic light are sought. For example- a woman who has loss of pin sensation in the left arm and leg. If she also had a left hemianopia, the localization is in the posterior portion of the right cerebral hemisphere (or two separate unrelated lesions). If, however, she had also had loss of pin sensation in the right face, right palatal weakness, right arm and leg ataxia, and nystagmus, the lesion would have to be in the right lateral medulla. Each symptom or sign should not be considered individually but should be used to try to define a locus or loci in the nervous system where these symptoms and signs would all localize.

The Topography of Local Symptoms is Also Helpful in Localization Headache in the forehead usually indicates a supratentorial lesion, while pain in the occiput or neck more often indicates an infratentorial posterior fossa localization. Pain and tenderness in the third thoracic vertebrae in a patient with paraparesis almost always suggests an extradural lesion at that location. Pain and tenderness in the left lower abdomen in a patient with left psoas and quadriceps muscle weakness makes it likely that the disease process affecting the left femoral nerve is located near the left psoas region.

The Neurologist generates and tests anatomical hypotheses during the history and examination much like he or she has tested disease mechanism hypotheses during the history. These anatomical hypotheses can usually be further refined or tested by neuroimaging or physiological tests.

The Modus Operandi- Sequential hypothesis generation and testing
Medical students and many trained physicians think of the clinical encounter as a passive, rote accumulation and recording of specific data items. They visualize a process much like filling out a checklist or completing a questionnaire. Then, after all the information is obtained, analysis and interpretation should lead to the correct answer.

Generation of clinical data should not be passive but should consist of very active thought from the minute the physician meets the patient. The experienced Neurologist, considering a particular anatomical localization or a disease mechanism, asks the patient questions that will test hypotheses he or she has in mind.

Patients are often naive about the workings of their own bodies, especially their nervous systems. Many patients with weakness or numbness in the left arm attribute the problem to a local process within that limb. They do not volunteer the occurrence of a temporary buckling of the left leg a month before, thinking the two problems completely unrelated. The same patient would be unlikely to relate the occurrence of transient monocular blindness in the right eye unless specifically asked about visual loss in the eyes. Patients often mentally divide up their bodies into the domains of various different specialists. Eye symptoms are brought to their ophthalmologists and "female problems" to their gynecologists. Patients don't know what to tell Neurologists. By generating anatomical hypotheses, the clinician-Neurologist asks the questions that will most likely lead to accurate localization. It takes more experience to know which questions to ask than to provide answers. Experience allows efficient interaction with the patient to ensure collection of all important relevant data.

Neurologists begin to generate hypotheses even before meeting the patient. These are based on information provided by the referring physician or layperson. After the initial statement of the problem by the patient, the physician begins to test these previous hypotheses and new ones suggested by the patient's initial remarks. After asking a series of penetrating questions, by following and listening carefully and digesting the responses, the Neurologist begins to organize and rate the hypotheses. When the history is completed, often while the patient is undressing, the physician should mentally list the active *what* hypotheses about the disease mechanism and *where* hypotheses about the clinical localization and be ready to test these during the examination.
As with the history, new, often unexpected findings are noted during the examination that alter previous hypotheses, change probabilities, or suggest new ideas. The findings can come from the general examination: a very high blood pressure, an unexpected black cutaneous lesion on the foot resembling a melanoma, clubbed fingers, or a loud carotid bruit or heart murmur. Unexpected neurological findings such as papilledema, an extensor plantar response, or tongue fasciculations also should change the physician's thinking and somehow must be integrated into the working hypotheses. During the examination, the alert clinician often returns to points in the history for clarification, and new examination findings may suggest historical questions not already asked. After the examination, the clinician should pause and again generate a modified list of diagnostic probabilities for both disease mechanism and localization. The Neurologist is now ready to plan laboratory investigations to test these hypotheses. Again, sequential modification of the diagnostic lists should follow each important laboratory result.

This systematic, sequential process of deductive reasoning is crucial if the neurologist is to (1) make the correct diagnosis, and (2) avoid serious diagnostic omissions. I teach this method to internists and medical residents because we believe it is equally applicable to diagnostic problems presented to internists. Unfortunately, the technique seems foreign to many of today's medical residents.

More Neurology is now practiced in out-patient settings than at any time in the past. After taking the history and examining a new patient, that individual and any accompany individuals expect the neurologist to intelligently explain what is wrong with the patient and what is to be done. If the physician has not been making and testing hypotheses during the encounter then or she will be totally unprepared to explain the problem at the end of the visit.

Probabilities and other diagnostic considerations
Probabilities are important. Rarely is a given diagnosis absolutely certain. Failure to think of and consider other conditions or localizations is the commonest reason for failure to diagnose. We all know physicians who invariably diagnose "zebras" instead of "horses" when hoofbeats are heard on the street. The probability of the presence of a given diagnosis is an important factor in planning investigations and treatment.
Another important factor is treatability of a disease. Even though a particular condition may be less likely than another, if it is remediable by treatment, then it is very important to carefully consider it before excluding it. Hypothyroidism, pernicious anemia, and insulin-secreting adenomas are unusual but potentially reversible causes of confusion, dementia, and ataxia.

Another consideration is the cost of missing a given diagnosis. By cost, I do not mean economic or legal, although these issues are becoming much more prominent today. We refer to morbidity and mortality. Subdural hematoma is not a common cause of hemiparesis but failure to diagnose can, and has, led to the death of patients who would have survived if treated quickly and effectively. Subarachnoid hemorrhage causes only a very small fraction of headaches, but failure to recognize the unusual instances can be disastrous.

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