I. TAKE HOME POINTS FOR THIS LECTURE

A. Subarachnoid hemorrhage (SAH) is fatal in 50% of cases, with >50% of survivors disabled by re-bleeding and vasospasm.

B. Headaches affect >50% of population, with chronic headache affecting >30% of women and >15% of men.

C. NEW Headaches are more likely due to underlying disorders (strokes, mass lesions, trauma, infections) than are chronic headaches.

D. Headaches can be effectively classified by their character, temporal features and triggering agents, but NOT by their severity.

E. Migraine has a central nervous system generator, and is treatable by use of serotonin agonists.

Headache is a very common complaint of patients seeing both primary care physicians and neurologists. Although headaches are usually "benign", they cause considerable morbidity and can be the harbinger of serious, even fatal disease. Subarachnoid hemorrhage stands alone as a worrisome cause of headache, and must be recognized immediately if the patient is to survive. Due to the commonness of headache, and the potential seriousness of the cause, a sophisticated approach to the headache patient is essential and not difficult to develop. Don’t get in the habit of blowing-off headache patients as stressed-out complainers; the headache is real and so is the potential for disaster. The goal of this chapter is to provide a framework within which you can evaluate headache patients.

II. SUBARACHNOID HEMORRHAGE

A. Terminology

1. SUBARACHNOID
   This refers to the space between the arachnoid membrane and the pia mater, the latter adhering directly to the brain parenchyma.

2. ANEURYSM
   Literally an arterial 'widening', the inherent structural flaw in the vessel wall at this site results in an increased potential for rupture.
   a) Congenital ("Berry")
      The most common type of aneurysm (present in 5-10% of the population), and the most frequent cause of subarachnoid hemorrhage, these balloon-shaped out-poochings of intracranial arteries occur at branch sites, presumably due to congenital flaws in the wall structure.
   b) Other
      Just so you know, there are several other kinds of intracranial arterial aneurysms that can be asymptomatic or can cause symptoms by means other than subarachnoid hemorrhage: "fusiform" longitudinal aneurysms; "Charcot-Bouchard" intraparenchymal aneurysms; "mycotic" aneurysms due to
septic emboli from the heart. So be precise when speaking of "aneurysms".

3. ARTERIOVENOUS MALFORMATION
A high flow anastamosis of arteries and veins bypassing the capillary bed. A more common cause of subarachnoid bleeding in the first two decades of life, often due to true aneurysms adjacent to the AVM.

4. VASOSPASM
Literally the spasm of cerebral arteries that occurs after a subarachnoid hemorrhage, somehow due to irritation of the arteries by the blood.

B. Anatomic Principles

1. COMPONENTS OF THE SUBARACHNOID SPACE
The subarachnoid space is continuous from the cerebral convexities to the cauda equina. It contains the cranial nerves and spinal roots as they cross the space, and large-caliber cerebral vessels, but is separated from the brain parenchyma by the pia mater. It is filled with the cerebrospinal fluid, which flows freely from cranial vault to lumbar sac.

2. LOCATION OF ANEURYSMS
Aneurysms occur at branch-sites of the major cerebral vessels at the base of the brain: middle cerebral, internal carotid, anterior communicating, anterior cerebral, posterior communicating. Bleeding tends to be most marked in the immediate vicinity of the aneurysm, allowing an educated guess as to the likely bleeding site when interpreting subarachnoid hemorrhage on a CT Scan.

C. Physiological Concepts

1. EFFECTS OF BLOOD PRESSURE
Elevated blood pressure is associated with aneurysmal rupture, and is a risk factor for re-bleeding in the hours or days following the initial hemorrhage. Low blood pressure is associated with increased risk of ischemia or infarction when vessels spasm (3-10 days after a bleed). Consequently, blood pressure must be kept "not too high and not too low" to avoid these conflicting devastating possibilities.
2. CHEMICAL "MENINGITIS"
Hemorrhage in the subarachnoid space produces irritation of the meninges and of the structures traversing the subarachnoid space, with resultant headache, stiff neck, "meningeal stretch signs", and possible cranial nerve palsies (e.g. III or VI nerve palsies with double vision). Unless damaged by the force of the bleed, the integrity of the pia mater prevents direct extension of blood into the brain parenchyma and thus prevents focal neurological signs. Some authors do not categorize subarachnoid hemorrhage as a stroke because of this absence of focal signs.

D. Clinical Features

1. DIAGNOSTIC PRINCIPLES
   a) Clinical Presentation
   The hallmark of subarachnoid hemorrhage is abrupt headache with altered mentation. The headache will be described below. Despite the lack of parenchymal damage, the toxic effects of blood in the subarachnoid space produce altered consciousness, with confusion and possibly coma. The absence of focal signs with marked deterioration of mental status has led some to describe subarachnoid hemorrhage as "sparing the soma but devastating the psyche". Look out for the abrupt onset of headache in association with this type of neurological change.

   b) Diagnostic evaluation
   The sine qua non of subarachnoid hemorrhage is blood in the cerebrospinal fluid. Blood can be identified either directly (drawn off by lumbar puncture) or by imaging it on CT Scan (NB: MRI Scans DO NOT show acute hemorrhage well, CT is the test of choice). The over-riding rule for identifying subarachnoid hemorrhage is to do as little as possible to make the diagnosis; any added stress, or any time wasted before treatment can result in re-bleeding and death.

2. THERAPEUTIC APPROACHES
Roughly 50% of patients die from a subarachnoid hemorrhage. Each re-bleed is associated with the same grim statistic. Surgical clipping of an aneurysm can be curative. The principle focus of treatment is to stabilize the patient, to prevent re-bleeding and to operate as soon as realistically possible. Blood pressure needs to be kept "not to high and not to low" prior to surgery, but can be artificially elevated after clipping to prevent the consequent ischemia that accompanies vasospasm. Medical management (with calcium channel blockers) can diminish, though not eliminate, vasospasm.
III. HEADACHE

Headache is the term used for various forms of discomfort perceived as originating in or on the head. As mentioned above, the clinician must be sophisticated in analyzing head pains to determine which are worrisome. Severity of pain is not helpful in diagnosing cause or type of headache. The goal of this section is to give you an approach to understanding cause and treatment of headache.

A. Terminology

1. SECONDARY HEADACHE DISORDERS

Headaches can be caused by various systemic or neurological conditions, and then are referred to as secondary headaches. This designation would include headaches due to trauma, hemorrhage, tumor, meningitis, hypercarbia, etc.

2. PRIMARY HEADACHE DISORDERS

Headaches are referred to as primary disorders if not associated with other recognizable neurological or systemic disease. Migraine and cluster headaches can be considered idiopathic or primary headaches.

B. Anatomic Principles

1. PAIN-SENSITIVE STRUCTURES OF THE HEAD

   a) Extracranial
      Skin, Muscle, Periosteum, Arteries, Paranasal sinuses

   b) Intracranial
      Dura mater, Large arteries, Venous sinuses

   The brain itself is not sensitive to painful stimuli. Consequently, intracerebral pathology produces headache only when it invades or otherwise affects (e.g. due to edema) pain-sensitive structures such as the dura mater or major vessels.

2. INTRACRANIAL REFERRED PAIN

   The ophthalmic division of the trigeminal nerve innervates the proximal portions of large intracranial vessels, and the meninges in the supratentorial region. Consequently, painful stimuli in these structures are referred to the eye or retro-orbital area that are also innervated by the first division of V. Rostral cervical roots innervate structures in the posterior fossa, so that noxious stimuli in this region are experienced as coming from the neck or posterior base of the skull.

C. Physiological Concepts

There appears to be a final common pathway for many headache disorders involving both the intracranial vessels and their innervation by the trigeminal nerve. The primary disorder may be in the CNS (as appears to happen in migraine), or may be uncertain, but almost always involves both the nerves and vessels. Hence the label "vascular" is inappropriate to differentiate headache types.
D. Clinical Features

1. DIAGNOSTIC PRINCIPLES

   a) New or Old
   Although cephalologists (as headache mavens can be called) distinguish many different headache syndromes, the most important headache classification system is to divide headaches into two types.

   HEADACHES ARE EITHER:

   1) NEW, or
   2) OLD.

   Chronicity signifies benignity. A NEW headache, whether a new type in a chronic headache patient or first-time headaches in a patient previously headache free, need to be taken very seriously. The first question in evaluating headache then should be: **IS IT NEW OR OLD?**

   b) Characteristics
   Severity of pain is extremely subjective, and not a useful diagnostic feature. So, what is ??

   CLINICALLY USEFUL FEATURES OF HEADACHE ARE:

   1) TEMPORAL PROFILE OF THE PAIN
   2) ASSOCIATED FEATURES
   3) LOCATION
   4) EXACERBATING AND RELIEVING FACTORS

2. THERAPEUTIC APPROACHES
   The treatment of headache disorders follows that of other painful conditions:

   a) remove any causative factors (i.e. treat the underlying disease)
   b) treat with disorder-specific medications
   c) for chronic benign headaches, use narcotics only as a last resort.

3. SPECIFIC DISORDERS SECONDARY TO UNDERLYING PATHOLOGY

   a) Subarachnoid hemorrhage
   **Time Course:** Abrupt, "instantaneous" peak (like an unexpected "thunderclap"), severe for hours, slowly diminishes over days.
   **Associated Features:** Change in level of consciousness, confused and afraid, meningeal stretch signs, possible cranial neuropathies.
   **Location:** global
   **Exacerbating and Relieving Factors:** Can occur with situations that elevate blood pressure: stress, exertion, sex
Clinical Picture: A NEW headache, described as "being hit over the head by a baseball bat", in a patient who is slightly sleepy.

b) Giant Cell Arteritis ("Temporal arteritis")
   Time Course: Evolves over months, can wax and wane but doesn't completely remit.
   Associated Features: Muscle aches, jaw claudication, weight loss, fever, ophthalmoplegia, elevated erythrocyte sedimentation rate.
   Location: Classically temporal, but can be anywhere. Tends to be superficial - "in the scalp"
   Exacerbating and Relieving Factors: Worsened by cold temperature, acutely relieved by steroids.
   Clinical Picture: This is a disorder of the elderly, almost all patients being >50y/o. It builds over weeks or months, but if not diagnosed and treated (with steroids) it can abruptly lead to permanent blindness or other irreversible cranial neuropathies.

c) Post-Lumbar Puncture Headache
   Time Course: Develops hours to days after lumbar puncture. Time course is totally dependent on position of patient.
   Associated Features: Non-specific
   Location: Global or sub-occipital.
   Exacerbating and Relieving Factors: Acutely sensitive to position. Gone when recumbent, occurs within seconds of sitting or standing.
   Clinical Picture: The patient is disabled by the headache because it occurs with an upright position. It is self-limited within 1-2 weeks, but can be treated by subdural injection of blood ("blood patch").

d) Other
   Increased intracranial pressure (from tumor, hemorrhage, or idiopathic), acute severe systemic hypertension, meningitis, sinusitis, dental disease and myriad other conditions are associated with headaches that are not always distinctive. Remember, however, that persistent focal pain indicates focal pathology.

4. PRIMARY HEADACHE DISORDERS

a) Migraine
   Time Course: Peaks in minutes to hours, lasts hours to days.
   Associated Features: Migrating symptoms of sensory disturbance (visual, tactile) that evolve and remit over 20 min., nausea/vomiting, photophobia.
   Location: Classically unilateral, though frequently global.
   Exacerbating and Relieving Factors: Can have discrete triggers (red wine, certain perfumes, too much sleep, caffeine withdrawal), relieved by sleep, frequently by aspirin.
Clinical Picture: Typically a long history of headaches, though they can worsen in severity. Migratory sensory symptoms evolving over 20 minutes, especially "scintillating scotoma", constitute an aura that is nearly pathognomonic for migraine. Headache often associated with an urge to sleep, and remits after sleeping. Treatment controls headache in the vast majority of migraineurs.

Pathophysiology: Etiology appears to involve a central neurogenic center involving serotonergic neurons, with subsequent slowly-spreading depression of cortical areas that accounts for the aura, and sensitization of blood vessel nociceptors.

b) "Tension"

Time Course: Waxing and waning for hours and days, if not weeks.

Clinical Picture: The distinction between migraine and tension headaches is often unclear. The vast majority of tension headache sufferers will report clear migraine symptoms some of the time. Everything is made worse by stress/tension, and muscle tension does not generate headache on its own. "Tension headache" should probably be treated as a form of chronic migraine.

c) Cluster

Time Course: Headaches recur daily during a bout, then cease for months until another cluster ensues. For each headache the onset to maximum pain is within 5-10 min., and pain is gone within 45-90 min.

Associated Features: Stuffy nose, tearing ipsilateral to pain.

Location: Unilateral temporal/retrobulbar.

Exacerbating and Relieving Factors: Exacerbated by alcohol ingestion, relieved by hyperventilation or inhaling 100% O₂.

Clinical Picture: Affects men>>women. Due to their brevity, each individual headache usually ends prior to seeking help. Most cluster cases can be controlled with medication.

IV. CASES – to be discussed in class.

V. REFERENCES


Headache, Raskin N.H., Churchill Livingstone, 1988

American Association for the Study of Headache Website:  
http://www.aash.org/education/

American Council for Headache Education Website:  
http://www.achenet.org/default.htm

AMA Health Insight Migraine Page
VI. SAMPLE EXAM QUESTION
A 29-year-old man, who had been perfectly healthy, came to the ER complaining of “the worst headache of his life”. On closer questioning, he mentioned that he had a history of “sinus headaches” that were very frequent, which typically cleared with sleep, and for which he was now taking Excedrin almost daily. The current headache had begun during the morning, shortly after a 20-minute period of bizarre flashing lights and blurred vision. The headache had slowly grown in severity during the course of the day, to the point where now, in late afternoon, the pain was unbearable. He could not tolerate having lights on in the exam room, was nauseated and actually vomited while in the ER.

1. Which of the following are the most likely underlying pathophysiological process affecting this patient?
   A. Increased pressure within the nasal sinuses.
   B. Vasospasm due to blood in the subarachnoid space.
   C. Increased pressure within cerebral ventricles
   D. Granulomatous inflammation of the extracranial vasculature
   E. A central nervous system generator that involves serotonergic neurons