Learning Objectives:
1. The learner will be able to differentiate primary from secondary headache disorders
2. The learner will accurately diagnose migraine as opposed to other painful disorders that cause eye pain
3. The learner will learn to differentiate the trigeminal autonomic cephalalgias
4. The learner will gain a more intimate knowledge of the International Classification of Headache Disorders system

CME Questions:
1. Which of the following must be present to make a diagnosis of migraine without aura?
   a. Unilateral location
   b. Throbbing
   c. Moderate or severe in intensity
   d. Nausea or light and sound sensitivity

2. Which of the following are allowed in the definition of tension type headache:
   a. Nausea
   b. Aura
   c. Photophobia
   d. Chemosis

3. Which of the following is not included in the category of “Trigeminal Autonomic Cephalalgia”
   a. SUNCT
   b. Chronic paroxysmal hemicrania
   c. Hemicrania continua
   d. Idiopathic stabbing headache

4. Unilateral eye pain associated with tearing and redness of the eye lasting 20 seconds and dissipating only to reoccur dozens of times a day is consistent with which of the following primary headache disorders?
   a. SUNCT
   b. Paroxysmal hemicrania
   c. Idiopathic stabbing headache
   d. Cluster headache

Keywords (Max 5):
1. Migraine
2. Tension type headache
3. Cluster headache
4. Trigeminal autonomic cephalalgias
5. Headache history
Introduction/Abstract:

Headache is one of the most common disorders presenting to the physicians office. Epidemiologic studies show that in a given year, the majority of people within the United States will have headache, and approximately 5% will seek medical attention. It is estimated that 25% of all new visits in a neurologists office is for headache. Over 90% of headaches are primary headache disorders that are they have no underlying secondary cause. As physicians, our main concern is finding the underlying disease or disorder that is causing headache. The primary headache disorders by definition have no significant abnormalities on their examination, nor relevant findings on neuroimaging. The key to making a diagnosis in the primary headache disorders taking a thorough history. Before 1988, the classification of headache and the diagnostic criteria for headaches was not uniform. Even the definition of migraine differed between the US and Europe. As a result of the need for clear headache definitions, and specifically migraine definitions, the International Headache Society formed a working group. In 1988, the International Headache Society released its first edition of a classification system for headache disorders. This classification system that has become the standard for headache diagnosis and has enhanced our ability to do multi-center headache research studies throughout the world. It has also facilitated medical practice, epidemiologic studies, and multinational clinical trials. The classification system is based on clinical consensus backed by data, experience, judgment, and compromise. The classification system has become the standard for headache diagnosis and clinical research and has become known as the International Classification for Headache Disorders (ICHD). The third edition of this classification is just been released in 2013 and is available online (http://www.ihsclassification.org/_downloads/mixed/International-Headache-Classification-III-ICHD-III-2013-Beta.pdf).

This classification will be utilized in the new ICD-10 coding system (note that the US still uses ICD-9, but will be going to ICD-10 in 2014 as mandated by CMS). The classification system separates out the primary and secondary headache disorders, as well as many other disorders that are at least somewhat linked to migraine.

The primary headache categories and coding is broken into the four sections:
1. Migraine
2. Tension-type headache (TTH)
3. Trigeminal autonomic cephalalgias
4. Other primary headaches

Body:

I. THE HISTORY

Since most headache patients have normal examinations, and since primary headache disorders by definition are normal interictally, the history becomes the most important tool for making a correct diagnosis. Patients often have more than one type of headache and it is important to have the time to sort out the nuances of the headache history. Family history is especially important in migraine, as 70% of migraine sufferers have first degree relatives with migraine.

A. Age at onset: Primary headache disorders often start in childhood and early adulthood. A childhood history of car sickness is very frequently seen in migraineurs. Also, a history of recurrent bouts of abdominal pain or vomiting in childhood may be the first manifestation of a migraine disorder. Later onset headaches have a more ominous differential diagnosis, and patients with onset of a new headache disorder after age 50 will need a more extensive evaluation, often including neuroimaging. It is important to start at the beginning of the headache disorder when headaches first started. Headache patterns often change over time, and new types of headache may appear. While open-ended questions are useful, one must drill down on specifics and ask specific questions. How has the headache changed? Are there new and different types of headache? Are the associated symptoms changing?

B. Location: Unilateral headaches are often associated with migraine, while cluster headaches are not only unilateral, but are always side locked; that is, always on the same side within each cluster period and in each subsequent cluster. Tension type headache on the other hand is typically bilateral.
Secondary headaches may also have very specific areas of pain. Supratentorial lesions often produce bi-frontal pain, while infratentorial lesions often cause occipital pain. Pain in and around the eye, which is of special importance to this group, may be related to diseases not only involving the orbit and the eye itself, but also may be referred pain from the cervical spine, the sinuses, the carotid artery, and the temporomandibular joint. However, in my practice, the most common cause of recurrent headache with pain in and around the eye is migraine.

C. Frequency and timing: Some headache disorders have fairly standard frequency; cluster headache is such an example. Most patients have one to three cluster headaches on a daily basis during the cluster episode, followed by a year or two of headache freedom. Cluster headache frequently occurs at the same time of day and night entraining to a circadian rhythm as well as a circannual rhythm. Migraine has common triggers that can be identified that lead to specific timing, for example, menstrual migraine. When dealing with the trigeminal autonomic cephalalgias (TAC), the symptoms may be very similar, but it is the duration of pain and frequency of attacks that differentiates them. Frequency is important not only to make a diagnosis, but also to determine degree of disability and thereby help select the most appropriate therapy. Headache specialists often use headache diaries to keep track of headache frequency, trigger factors, and use of medication. It is important to sort out clearly identifiable timing factors that will help in making a specific diagnosis and avoiding an unnecessary work-up. The one related red flag to always consider is a headache that is increasing in frequency and or severity. This may be a harbinger of a secondary headache disorder.

D. Onset, Duration, Character, and Severity: Headache duration is one of the more important factors in sorting out the primary headache disorders. Pain intensity is also very important not only for diagnosis, but to determine best therapy. Patients with frequent but very mild headaches will often not require the same intensity of treatment than the much less frequent but very severe headache sufferer may require. It is helpful to have the patient use a pain scale. The most commonly used scale is a 1 to 10 scale. It’s always interesting however, when a patient is asked this question and responds that all of the headaches are a 12, while sitting in front of you with their “12” headache, smiling, engaging, and looking well. Headaches with sudden onset and peaking over seconds are termed thunderclap headaches. While this may represent primary thunderclap headache, one must assume a potentially serious etiology such as aneurysmal rupture or venous sinus thrombosis, and evaluate on an emergent basis leaving primary thunderclap headache as a diagnosis of exclusion. While severity is certainly a factor when considering the etiology of a headache, it’s important to remember that the quality of migraine is often severe, and pain severity does not necessarily equate to a more ominous diagnosis. More serious causes of headache such as brain tumor or AVM are often associated with mild nonspecific headaches types of headache. The overall intensity of the headache is not directly correlated with the seriousness of the underlying cause.

Headache duration is very helpful in determining the type of headache disorder a patient has, especially when dealing with the primary headache disorders. Headaches lasting less than 4 hours are termed the short-lasting headaches, and are in a different bucket that the longer lasting primary headaches, i.e. Migraine, and Tension Type Headache. Migraine by definition lasts 4 to 72 hours. Cluster headaches typically last 30-120 minutes, while SUNCT, one of the TACs has episodes of pain that last 5-120 seconds.

E. Associated symptoms: The type of associated symptoms that occur with each headache disorder can be very important in limiting the differential diagnosis. Some rules to live by:

a. Nausea may be seen with any severe headache, be it primary or secondary, but usually points to migraine. Tension type headache by definition is not associated with nausea
b. Vertigo is a common symptom seen with migraine
c. While visual aura is typical of migraine, there are patients with cluster headache who may have aura
d. Headaches starting in the neck or shoulder may still be migraine
e. Nasal congestion may be seen with migraine and does not indicate “sinus headache”
f. Migraine visual aura may rarely be secondary to an irritative lesion such as an occipital lobe tumor or AVM in which case the visual disturbance is always on the same side and a visual field defect is usually present
g. Polyuria may be associated with migraine, but not other headache disorders
h. Mood disorders and cognitive dysfunction are commonly seen both as a prodrome to migraine and with migraine
i. Lying down may exacerbate migraine pain and some patients are more comfortable sitting quietly in a chair
j. Headaches worsened by standing and much improved by laying down suggests intracranial hypotension
k. Side locked headaches are common in migraine, and while they are red flags in some textbooks, there is no evidence of increased incidence of secondary headache

II. THE EXAMINATION

Patients referred to neuro-ophthalmologist will receive a thorough examination of the eyes as well as a neurologic examination. The emphasis may change according to the specialty training of the examiner, but there are several important features to review with regards to headache disorders.

a. Palpate (gently) and listen to the carotid artery
b. Palpate the temporal arteries at the ear and forehead looking for thrombosis, nodules, absent pulse, and tenderness
c. Palpate the sinuses, globes, trochlear apparatus, temporal regions, and temporomandibular joint
d. Palpate the sub-occipital region, especially when the patient has chronic unexplained retro-orbital pain looking for a trigger point
e. Listen for a cranial bruit
f. Take the blood pressure
g. Examine all three distributions of the trigeminal nerve including muscles of mastication

III. THE PRIMARY HEADACHE DISORDERS:

A. MIGRAINE

Migraine is a common disorder with a similar epidemiology throughout the world. In the US, the prevalence of migraine is 8% in men and 18% in women. In a survey of neurologists attending a headache course, the 1-year and lifetime prevalence of migraine in the 220 respondents were as follows: male neurologists, 34.7%, 46.6%; male headache specialists, 59.3%, 71.9%; female neurologists, 58.1%, 62.8%; and female headache specialists, 74.1%, 81.5%. 4% of the population has chronic daily headache, and 1% have chronic migraine (15 or more headache days a month). 20% of migraine sufferers have migraine with aura, and 80% have migraine without aura. Migraine is a diagnosis of inclusion. The IHS criteria for the diagnosis of migraine are relatively simple and straightforward. The diagnosis of migraine is made on the basis of history alone, as there are no objective findings on examination or any laboratory tests that confirm the diagnosis. Studies have shown that the majority of patients who visit a physician with the chief complaint of headache have migraine. In a study done in England, Europe, and the United States, patients seen in a primary care setting for a chief complaint of headache were evaluated and asked to keep a diary of their subsequent headaches. Headache specialists reviewed these diaries and the headaches were classified. In this study, 76% of patients met the International Headache Society criteria of migraine, 18% satisfied the criteria for probable migraine, 3% had tension-type headache, and 3% had other types of headache. In another interesting observational study of patients seen in an emergency department with a primary diagnosis of headache, only 32% received a diagnosis of migraine while 59% received a diagnosis of “headache not specified.” When the charts were reviewed and the patients interviewed, 95% of the patients seen for primary complaint of headache in the emergency department met the IHS criteria for migraine.

Other features that strongly suggest migraine include predictable timing around menses, stereotyped premonitory symptoms, characteristic triggers, and improvement with sleep especially in children, positive family history, and smell sensitivity. Many patients have premonitory symptoms including yawning, food cravings, and behavioral changes, which may precede their headache by hours to a day or more. A positive family history is seen in up to 70% of migraine patients if the patient is correctly questioned. Sometimes it is difficult to tease out the history, and leading questions may provide more information. Rather than just asking if there is a positive family history of migraine, one might ask if anybody in the family has sick headaches, headaches that put them in bed or headaches that are disabling. Many patients also have characteristic triggers for their migraine. Certain foods such as nuts, citrus fruits, aged cheeses, and cured meats may be triggers for some patients. Alcohol, especially red wine, is a potent migraine trigger. Other triggers include sleeping longer than usual in the morning, sudden increase or
reduction of stress, and missing meals. So-called “let down” migraine is especially troubling to patients. These occur in migraine patients who have a reduction in stress due to vacation, weekends, or after major life events.

Although some patients with migraine never receive a diagnosis of migraine, a significant number are misdiagnosed. Of the 52% of patients with migraine who’ve not been diagnosed as having migraine in Lipton’s American Migraine study, 32% of those patients had been diagnosed as having tension-type headache, and 42% of those patients had been diagnosed as having sinus headache. They typically receive the diagnosis of tension headache because the pain starts in the neck, the headache is bilateral, or the headache is associated with stress. While a diagnosis of tension-type headache is often given in a patient who actually has migraine, the most common diagnosis mistakenly given to a migraine patient is “sinus headache.” Acute sinusitis may be associated with headache and facial pain, but chronic allergic sinus disease, as well as sinus and nasal congestion, are not associated with severe and disabling headaches. Patients are often given a diagnosis of sinus headache because their headaches come on with weather change, and the location of the headache is typically behind the eyes and in the forehead, which patients interpret as coming from the sinuses. In an interview study of 177 patients with definite migraine headaches, 46% of patients had at least one autonomic symptom during their migraine attack; 46% had both nasal and ocular symptoms; 14% had nasal symptoms alone; and 41% had ocular symptoms alone. The ocular symptoms included tearing, redness, and eyelid edema. In an interesting study done by Cady, 24 patients were recruit through a newspaper advertisement seeking patients who believed they had sinus headache and had never been given a diagnosis of migraine. Nearly 90% of these patients met the ICHD-2 criteria for migraine. In addition, they responded to sumatriptan in the same manner as a typical migraine patient. Despite the frequency with which a diagnosis of sinus headache is given, the data and clinical experience suggests that the vast majority of patients who are given a diagnosis of sinus headache, or believe they have sinus headache, actually have migraine.

**Migraine without aura: ICHD 1.1**

**Description:**
Recurrent headache disorder manifesting in attacks lasting 4-72 hours. Typical characteristics of the headache are unilateral location, pulsating quality, moderate or severe intensity, aggravation by routine physical activity and association with nausea and/or photophobia and phonophobia.

**Diagnostic Criteria**
A. At least five attacks, fulfilling criteria B–D
B. Headache attacks lasting 4-72 hours (untreated or unsuccessfully treated)
C. Headache has at least two of the following four characteristics:
   1. unilateral location
   2. pulsating quality
   3. moderate or severe pain intensity
   4. aggravation by or causing avoidance of routine physical activity (e.g. walking or climbing stairs)
D. During headache at least one of the following:
   1. nausea and/or vomiting
   2. photophobia and phonophobia

**Migraine with Aura: ICHD 1.2**

**Description:**
Recurrent attacks, lasting minutes, of unilateral fully reversible visual, sensory or other central nervous system symptoms that usually develop gradually and are usually followed by headache and associated migraine symptoms.

**Diagnostic Criteria:**
A. At least two attacks fulfilling criteria B and C
B. One or more of the following fully reversible aura symptoms:
   1. visual
   2. sensory
3. speech and/or language
4. motor
5. brainstem
6. retinal

C. At least two of the following four characteristics:
   1. at least one aura symptom spreads gradually over 5 minutes, and/or two or more symptoms occur in succession
   2. each individual aura symptom lasts 5-60 minutes
   3. at least one aura symptom is unilateral
   4. the aura is accompanied, or followed within 60 minutes, by headache

D. Not better accounted for by another ICHD-3 diagnosis, and transient ischemic attack has been ruled out

B. TENSION TYPE HEADACHE: Tension type headache (TTHA) is the most common type of headache, and it is estimated that lifetime prevalence is 90%. TTHA is everything that is “not migraine”. It is typically mild to moderate in intensity, bilateral, non-throbbing, not increased with exertion, and not associated with nausea. Due to the relatively mild nature of these headaches, patients with episodic TTHA typically do not seek medical care and tend to self-treat with over the counter medications. In the past it was believed that most chronic daily headache was TTHA, but we now understand that most of these patients more likely have chronic migraine. Prevalence of chronic TTHA is estimated at 2%. It would be unusual for patients with TTHA to have significant eye pain, or end up in the office of a neuro-ophthalmologist. However, many patients with chronic daily headache will have associated pain in the frontal and orbital regions. TTHA is often associated with trigger points, pericranial tenderness, lower pain threshold to scalp pressure, and head forward posture.

A study by Schwartz and coworkers estimated the overall prevalence of episodic tension-type headache at the following:

- 38.3% of the population
- Females age 30–39 years, approximately 47%
- African-American descent overall, 22.8%
- Increased with increasing levels of education
- Graduate school level education, approximately 50%

Tension Type Headache ICHD 2.X

Description:
Episodes of headache, typically bilateral, pressing or tightening in quality and of mild to moderate intensity, lasting minutes to days. The pain does not worsen with routine physical activity and is not associated with nausea, but photophobia or phonophobia may be present.

Diagnostic criteria:
A. At least 10 episodes of headache fulfilling criteria B
   - Infrequent (<1d/month)
   - Frequent (1-14d/month)
   - Chronic (>14d/mo)
B. Lasting from 30 minutes to 7 days
C. At least two of the following four characteristics:
   1. bilateral location
   2. pressing or tightening (non-pulsating) quality
   3. mild or moderate intensity
   4. not aggravated by routine physical activity such as walking or climbing stairs
D. Both of the following:
   1. no nausea or vomiting
   2. no more than one of photophobia or phonophobia

C. THE TRIGEMINAL AUTONOMIC CEPHALALGIAS

1. Cluster Headache: Cluster headache is a relatively rare primary headache disorder, but the
symptoms are so stereotyped that it is usually not difficult to diagnose to the informed care provider. It may be the most painful of all of the primary headache disorders. Known in the past by a variety of monikers (migrainous neuralgia, histamine cephalgia, petrosal neuralgia, sphenopalatine neuralgia, vidian neuralgia, and Sluder’s neuralgia), it was classified as a variety of migraine until the 1988 IHS classification, when it was included in the trigeminal autonomic cephalalgias. The best early description was made by Horton at the Mayo in 1939, at which time they treated with histamine desensitization and the syndrome became known as Horton’s histamine cephalgia ⁹. In 1952, E.C. Kunkle named the syndrome “cluster headache” and this is the name that has been adopted by the Ad Hoc Committee in 1962 and the World Federation of Neurology in 1969 ¹⁰. The cluster headache itself often referred to as an attack, typically lasts 60-90 minutes, while the cluster period, which is the time in which the patient has recurrent headaches typically, lasts 4-8 weeks. Most patients have episodic cluster with cluster periods occurring once a year or every other year, often at the same time of year (circannual). Patients with chronic cluster, have headaches on a nearly daily basis with no prolonged headache free interval. There are some patients with typical cluster headache who also have typical lancinating facial pain consistent with trigeminal neuralgia and this has been referred to as “cluster-tic”. The term “cluster migraine” was coined by Medina and Diamond ¹¹ in 1977 to describe patients who have both cluster headache and migraine headache. However it is a term that is misused to describe patients with migraine who have “clusters” of migraine followed by periods with no migraine. The term cluster migraine should be avoided. The usual attack consists of the rapid onset of unilateral periorbital headache that builds to a peak in about 10 to 15 minutes and lasts for approximately 30 to 180 minutes. During a cluster attack, patients are agitated and prefer to pace in contradistinction to migraine where they want to be still. Attacks are characterized by excruciating pain that is regarded by sufferers as the worst pain that they have ever experienced. The pain is associated with ipsilateral cranial parasympathetic over activity with may cause one or more of the following: lacrimation, conjunctival injection and nasal stuffiness or rhinorrhea, lid edema, and an ipsilateral partial (third order neuron) Horner syndrome with ptosis and miosis. The headache is unilateral. In order of decreasing frequency, common sites of pain are: orbital, retro-orbital, temporal, supraorbital, and infraorbital. The number of attacks per day varies, usually from 1 to 3, but the International Headache Society classification allows from 1 every second day to 8 per day within the definition of cluster headache. Migrainous features, such as nausea, photophobia, and phonophobia, can be present in as many as half of cluster headache cases ¹². Moreover, between 6% ¹³ and 14% ¹² of patients report typical migraine aura with their cluster headache. Cluster headache involves dysfunction of central nervous system elements concerned with pain control and with links to circadian and circannual mechanisms. Research suggests the hypothalamus is intricately involved in the pathophysiology of cluster headache.

**Cluster Headache ICHD 3.1**

**Description:**
Attacks of severe, strictly unilateral pain, which is orbital, supraorbital, temporal or in any combination of these sites, lasting 15–180 minutes and occurring from once every other day to eight times a day. The pain is associated with ipsilateral conjunctival injection, lacrimation, nasal congestion, rhinorrhea, forehead and facial sweating, miosis, ptosis and/or eyelid edema, and/or with restlessness or agitation.

**Diagnostic criteria:**
A. At least five attacks fulfilling criteria B–D
B. Severe or very severe unilateral orbital, supraorbital and/or temporal pain lasting 15–180 minutes (when untreated)
C. Either or both of the following:
   1. at least one of the following symptoms or signs, ipsilateral to the headache:
      a) conjunctival injection and/or lacrimation
      b) nasal congestion and/or rhinorrhea
      c) eyelid edema
      d) forehead and facial sweating
      e) forehead and facial flushing
      f) sensation of fullness in the ear
      g) miosis and/or ptosis
   2. a sense of restlessness or agitation
D. Attacks have a frequency between one every other day and eight per day for more than half of the time when the disorder is active
2. **Paroxysmal Hemicrania**: This is a rare headache disorder, seen much less commonly than cluster headache. It can be episodic or chronic, but the chronic form is more typical. Some patients start with the episodic form and over time transform into the more typical chronic form. Chronic paroxysmal hemicrania (CPH), was first described in 1974 by Sjaastad and Dale 14. In 1976, the term CPH was proposed by Sjaastad 15 on the basis of the first 2 patients, who had daily solitary, limited attacks (i.e., paroxysmal) of unilateral headache that did not shift sides (i.e., hemicrania). The clinical diagnosis is based on attacks of severe, unilateral orbital, supraorbital, and/or temporal pain, always on the same side, lasting 2-45 minutes. Pain is associated with at least 1 of the following signs/symptoms on the symptomatic side:

- conjunctival injection
- lacrimation
- nasal congestion
- rhinorrhea
- ptosis
- eyelid edema

Most patients who have CPH exhibit lacrimation (62%), followed by nasal congestion (42%), conjunctival injection and rhinorrhea (36%), and ptosis (33%) 16. There may be mild meiosis, but not a typical sympathetic defect as one sees in cluster headache. In some patients CPH attacks can be triggered by a variety of triggers such as rotating the neck or flexing the head to the side of the headaches or by applying external pressure to the transverse processes of C4 to C5 or the C2 nerve root on the symptomatic side. The attack frequency typically ranges from 10-20 attacks daily, but may be more. Attacks usually last 2-25 minutes, but they may last as long as 60 minutes. By definition, this is one of the Indomethacin responsive headache disorders, and in order to make the diagnosis, the patient must respond to Indomethacin. There are case reports of paroxysmal hemicrania like headaches that are secondary to parasellar disease, orbital pathology, and vascular disorders. These patients should be imaged with contrast enhanced MRI.

The etiology of this headache disorder is unknown, and there is no genetic component. Functional imaging studies have demonstrated involvement of the hypothalamus. 17. Average age of onset is 34, and this entity may be seen in children. There is a 3:1 female predominance.

**Paroxysmal Hemicrania ICHD 3.2**

**Description:**

Attacks of severe, strictly unilateral pain which is orbital, supraorbital, temporal or in any combination of these sites, lasting 2–30 minutes and occurring several or many times a day. The attacks are associated with ipsilateral conjunctival injection, lacrimation, nasal congestion, rhinorrhea, forehead and facial sweating, miosis, ptosis and/or eyelid edema. They respond absolutely to Indomethacin.

**Diagnostic criteria:**

A. At least 20 attacks fulfilling criteria B-E
B. Severe unilateral orbital, supraorbital and/or temporal pain lasting 2–30 minutes
C. At least one of the following symptoms or signs, ipsilateral to the pain:
   1. conjunctival injection and/or lacrimation
   2. nasal congestion and/or rhinorrhea
   3. eyelid edema
   4. forehead and facial sweating
   5. forehead and facial flushing
   6. sensation of fullness in the ear
   7. miosis and/or ptosis
D. Attacks have a frequency above five per day for more than half of the time
E. Attacks are prevented absolutely by therapeutic doses of Indomethacin
F. Not better accounted for by another ICHD-3 diagnosis.

3. **SUNA (Short-lasting Unilateral Neuralgiform headache with cranial autonomic features) and SUNCT: (Short-lasting Unilateral Neuralgiform headache attacks with Conjunctival injection and Tearing syndrome)**

The SUNCT syndrome was first described by Sjaastad and colleagues in 1978 18. The description of the
complete syndrome came in 1989 again by Sjaastad. In the ICHD-3 classification, SUNCT and SUNA are both a subclass of “short-lasting unilateral neuralgiform headache attacks”. By definition, SUNCT patients always have both conjunctival injection and tearing, and the symptoms are a prominent part of the syndrome. Patients with SUNA have at least one autonomic feature, and the autonomic symptoms tend to be much less intense than in SUNCT. These symptoms include:

- conjunctival injection and/or lacrimation
- nasal congestion and/or rhinorrhea
- eyelid edema
- forehead and facial sweating
- forehead and facial flushing
- sensation of fullness in the ear
- miosis and/or ptosis

SUNCT is much more common in men than women with a M:F ratio of 17:2. The typical age of onset is between 40 and 70 years and the mean age of onset is 51 years. Pain severity is normally moderate to severe, unlike cluster headache, which is always severe. Pain duration is extremely short, usually lasting between 5 and 240 seconds, with an average duration of 10 to 60 second. The brevity of SUNCT sets it apart from all of the other primary headache disorders except for cranial neuralgias such as trigeminal neuralgia and stabbing headache pains. Pain onset is typically abrupt, with maximum intensity being reached in a few seconds. The pain normally plateaus at a maximum intensity for several seconds and then quickly abates. Mean attack frequency is 28 attacks per day, but may range form 1-80 with a tendency to have frequent attacks followed by a period of remission. SUNCT syndrome is an episodic disorder that presents in a relapsing and remitting pattern. Each symptomatic period can last from several days to several months, and a person who has SUNCT syndrome typically has one to two symptomatic periods a year (20). By definition, patients with SUNCT have conjunctival injection and tearing, but many may have other autonomic features. Ipsilateral rhinorrhea or nasal congestion occurs in 2/3 of patients. Other less common associated symptoms include eyelid edema, narrowed palpebral fissure, facial erythema, and photophobia. Typically, conjunctival injection and eye tearing start within 1 to 2 seconds of pain onset and remain until the head pain ceases. While SUNCT can occur spontaneously, many sufferers have triggers including chewing, nose blowing, coughing, forehead touching, eyelid squeezing, neck movements (rotation, extension, and flexion), and eating ice cream. SUNCT and SUNA are not Indomethacin responsive like most of the other trigeminal autonomic cephalalgias. There is some evidence that microvascular compression of the trigeminal nerve may be causative in some patients as microvascular decompression has been successful in providing complete remission is greater than 50% of patients.

Short-lasting unilateral neuralgiform headache attacks ICHD 3.3

Description:
Attacks of moderate or severe, strictly unilateral head pain lasting seconds to minutes, occurring at least once a day and usually associated with prominent lacrimation and redness of the ipsilateral eye.

Diagnostic criteria:
A. At least 20 attacks fulfilling criteria B–D
B. Moderate or severe unilateral head pain, with orbital, supraorbital, temporal and/or other trigeminal distribution, lasting for 1–600 seconds and occurring as single stabs, series of stabs or in a sawtooth pattern
C. At least one of the following cranial autonomic symptoms or signs, ipsilateral to the pain:
   1. conjunctival injection and/or lacrimation
   2. nasal congestion and/or rhinorrhea
   3. eyelid edema
   4. forehead and facial sweating
   5. forehead and facial flushing
   6. sensation of fullness in the ear
   7. miosis and/or ptosis
D. Attacks have a frequency of at least one a day for more than half of the time when the disorder is active

Short-lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing (SUNCT)
Diagnostic criteria:
A. Attacks fulfilling criteria for 3.3 Short-lasting unilateral neuralgiform headache attacks
B. Both conjunctival injection and lacrimation (tearing).

Short lasting unilateral neuralgiform headache attacks with cranial autonomic symptoms (SUNA)
Diagnostic criteria:
A. Attacks fulfilling criteria for 3.3 Short-lasting unilateral neuralgiform headache attacks, and criterion B below
B. Only one or neither of conjunctival injection and lacrimation (tearing).

4. Hemicrania Continua (HC): The term hemicrania continua was first introduced in 1984 by Sjaastad and Spierings. HC is a unilateral headache with varying intensity. It is continuous for months and years at a time and there are paroxysmal episodes of worsening pain lasting 20 minutes to 48 hours associated with autonomic features. These include:
- conjunctival injection
- lacrimation
- nasal congestion
- rhinorrhea
- eyelid edema
- forehead and facial sweating
- forehead and facial flushing
- sensation of fullness in the ear
- miosis and/or ptosis.

Patients often become agitated and irritable. During these exacerbations, there may be typical migraine symptoms as well, including nausea, vomiting, photophobia, and phonophobia. Some patients complain of a foreign body sensation in the eye, and others have typical ice-pick pains, especially with exacerbations. In Cittadini and Goadsby’s study of 39 patients, the pain was reported in the following locations: temporal, 82 percent; orbital, 67 percent; frontal, 64 percent; retro-orbital, 59 percent; occipital and parietal, 54 percent; vertex and periorbital, 51 percent; neck, 33 percent; maxillary and ear, 30 percent; upper teeth, 20 percent; shoulder, 18 percent; nose, 15 percent; jaw, 15 percent; eyebrow and lower teeth, 10 percent; retro-auricular area, eight percent; and upper and lower gum, two percent. HC can be labeled chronic when daily and continuous without pain-free periods for a minimum of one year and episodic when there are pain-free intervals of at least a day without treatment. HC is one of the Indomethacin responsive headache disorders, and like CPH, absolutely responds to Indomethacin at appropriate doses.

Hemicrania continua ICHD 3.4
Description:
Persistent, strictly unilateral headache, associated with conjunctival injection, lacrimation, nasal congestion, rhinorrhea, forehead and facial sweating, miosis, ptosis and/or eyelid edema, and/or with restlessness or agitation. The headache is absolutely sensitive to Indomethacin.

Diagnostic criteria:
A. Unilateral headache fulfilling criteria B-D
B. Present for >3 months, with exacerbations of moderate or greater intensity
C. Either or both of the following:
   1. at least one of the following symptoms or signs, ipsilateral to the headache:
      a) conjunctival injection and/or lacrimation
b) nasal congestion and/or rhinorrhea
c) eyelid edema
d) forehead and facial sweating
e) forehead and facial flushing
f) sensation of fullness in the ear
g) miosis and/or ptosis

2. a sense of restlessness or agitation, or aggravation of the pain by movement

D. Responds absolutely to therapeutic doses of Indomethacin

D. PRIMARY STABBING HEADACHE (PSH): This entity is associated with brief sharp recurrent pains that occur in the head and face, mostly in the V1 distribution. This condition has been known in the past as ice-pick pains, jabs and jolts, needle-in-the-eye syndrome, and ophthalmodynia periodica. It seen more commonly in migraine patients, but is quite common in the general population with one study suggesting a prevalence rate as high as 35% with average age of onset of 28 and a female predominance.

Raskin compared the incidence and the clinical characteristics of PSH in 100 migraineurs and 100 control subjects. PSH was found in 42% of migraineurs versus only 3% of healthy controls. These sharp stabbing or ice pick pains are often felt in the eye and periorbital region prompting ophthalmologic or neuro-ophthalmologic evaluation. The examination is always normal. Studies show 80% of stabs last 3 seconds or less. Attack frequency is generally low, with one or a few per day. In rare cases, stabs occur repetitively over days. It may move from one area to another on the head and face, in either the same or the opposite hemicranium: in only one-third of patients it has a fixed location. A few patients have accompanying symptoms, but not including cranial autonomic symptoms. Presence of any autonomic symptoms help to differentiate PSH from Short-lasting unilateral neuralgiform headache attacks. There is some speculation that PSH may be associated with venous sinus stenosis. The headache disorder is usually responsive to Indomethacin, but treatment is rarely necessary.

Primary stabbing headache ICHD 4.7

Description:
Transient and localized stabs of pain in the head that occur spontaneously in the absence of organic disease of underlying structures or of the cranial nerves.

Diagnostic criteria:
A. Head pain occurring spontaneously as a single stab or series of stabs and fulfilling criteria B–D
B. Each stab lasts for up to a few seconds
C. Stabs recur with irregular frequency, from one to many per day
D. No cranial autonomic symptoms

E. TRIGEMINAL NEURALGIA (TGN): TGN is not typically included as a primary headache disorder, but rather it is classified as a neuralgia. Nicolas André coined the term tic douloureux in 1756 in a book, Observations pratiques sur les maladies de l'urethre et sur plusieurs faits convulsifs. There is a rich history of descriptions of the condition, which likely represents TGN which can be found in the writings of Galen, Aretaeus of Cappadocia (born circa AD 81), and in the 11th century by Avicenna ("tortura oris")

John Fothergill was the first to give a full and accurate description of this condition in a paper titled "On a Painful Affliction of the Face," which he presented to the medical society of London in 1773. It was called Fothergill’s disease for the next 150 years. Average age of onset is about 60 and it is more common in women than men in a ratio of 2.5:1. While vascular compression is the most common cause of TGN, posterior fossa tumors, and multiple sclerosis should be considered, especially in the younger patient. As many as 15% of trigeminal cases are due to secondary causes so neuroimaging is often recommended. Red flags in trigeminal neuralgia include sensory loss and bilateral involvement.

TGN typically presents with brief stabbing unilateral facial pain, more often on the right side of the face. Patients may only have one or two attacks a day, but some may have hundreds. Typical triggers include: chewing, talking, smiling, drinking cold or hot fluids, touching a specific pain trigger point, shaving.
brushing teeth, blowing the nose or a draft. Patients can usually localize their pain precisely and the pain usually stays in the distribution of one division of the trigeminal nerve or occurs at the border zone of two nerves. The most common location is the corner of the mouth with radiation to the angle of the jaw. The ophthalmic distribution is involved in less than 5% of cases, so ophthalmologists are usually not involved in the early diagnosis. The pain is typically severe, paroxysmal, and lancinating. It is described as like an electrical shock which lasts 1-2 seconds but may continue for 20-30 seconds leaving the patient with a deep aching. Many patients have a refractory period and become pain free between paroxysms of pain.

**Trigeminal Neuralgia ICHD 13.1**

**Description:**
Trigeminal neuralgia developing without apparent cause other than neurovascular compression.

**Diagnostic criteria:**
A. At least three attacks of unilateral facial pain fulfilling criteria B and C
B. Occurring in one or more divisions of the trigeminal nerve, with no radiation beyond the trigeminal distribution
C. Pain has at least three of the following four characteristics:
   1. recurring in paroxysmal attacks lasting from a fraction of a second to 2 minutes
   2. severe intensity
   3. electric shock-like, shooting, stabbing or sharp in quality
   4. precipitated by innocuous stimuli to the affected side of the face
D. No clinically evident neurological deficit

**CME Answers:**
1. d)
2. c)
3. d)
4. a)

**References:**
29. Fothergill J. Of a painful affection of the face. Medical observations and inquiries by a society of physicians. London, 1773;5:129–142 [This publication was funded privately by Fothergill from 1771–1776. It is included in Complete Works of John Fothergill, a copy in library of RCP London.]