ABSTRACT:

Headache is a symptom experienced by all some time or other. It is caused by multitude of ailments of the body. The symptom is analyzed in reference to clinical history to arrive at diagnosis.

Key words: Secondary and Primary headaches -Trigeminal Vascular System- Cranial Autonomic Nervous System- Pain Sensitive Modulation.

Headaches are universal experience with lifetime prevalence 99%. 9% of adults see primary care physicians and 83% self medicate. It may be as simple as relief with rest or a cup of coffee, to uncommon life threatening situation. In most of the cases a careful history is adequate for precise diagnosis and hence understanding anatomy, physiology and pharmacology concerned with headache helps in better interpretation of history.

There are about 300 different conditions causing a single symptom of headache, with limited number of pain sensitive structures giving rise to similar headaches. Longer structure brain Parenchyma is insensitive to pain.

The pain location may be at the source like pain due to maxillary or frontal sinusitis or away from location – referred pains. For example, supratentorial structures are innervated by the ophthalmic division of trigeminal nerve, and the infratentorial or posterior fossa structures are supplied by C2 and C3. Thus a cerebellar hemisphere lesion generally refers pain posteriorly and occipital lobe lesion refers pain anteriorly.

ANATOMY, PHYSIOLOGY AND PATHOGENESIS OF HEADACHE

Pain sensitive structures that can cause headache are

Intracranial structures

Dura near vessels, Cranial nerves V, VII, IX, X, Circle of Willis and proximal continuations, Meningeal arteries, Large veins in the brain and dura

External to the skull

Scalp and neck muscles, Cervical nerves and roots, Cutaneous nerves and skin

Mucosa of the paranasal sinuses, Teeth, External carotid arteries and branches.

Two systems are involved in causing Headache syndromes1,2.

1. Trigeminal vascular system consists of innervations of large cranial vessels and dura mater by the Trigeminal nerve, containing substance P and calcitonin gene –related peptide (CGRP) which can be released when the trigeminal ganglion is stimulated.

2. Cranial Autonomic system causing lacrimation, nasal congestion and vascular changes. Pharmacological data suggest involvement of 5- Hydroxy tryptamine- serotonin and dopamine in migraine. 5 –hydroxy tryptamine 1B/1D receptor agonist or Dopamine receptor antagonists have anti –migraine efficacy.

Common causes of Headache

<table>
<thead>
<tr>
<th>Type</th>
<th>Primary Headache</th>
<th>Secondary Headache</th>
</tr>
</thead>
<tbody>
<tr>
<td>Migraine</td>
<td>16%</td>
<td>Systemic infection</td>
</tr>
<tr>
<td>Tension type</td>
<td>69%</td>
<td>Head injury</td>
</tr>
<tr>
<td>Cluster</td>
<td>0.1%</td>
<td>Vascular disorders</td>
</tr>
<tr>
<td>Idiopathic Stabbing</td>
<td>2%</td>
<td>Sub arachnoid Hemorrhage</td>
</tr>
<tr>
<td>Exertional</td>
<td>1%</td>
<td>Brain Tumour</td>
</tr>
</tbody>
</table>

Source: After J.Olesen et al . The headaches philadelphia, Lippincott, Williams and Wilkins, 2005

From the table it is evident that the most common causes of headache are Tension type of Headache and Systemic Infections. The management of either group is diagonally opposite. Hence the importance of the precise diagnosis which is basically dependent on history, once the neuro imaging is negative or normal.

Elements of headache history

Temporal profile

Age of onset, Frequency, Time of day, Duration, Recurrence

Headache features

Location, Quality of pain, Severity of pain

Associated symptoms and signs
Before headache, During headache, After headache

Aggravating and precipitating factors
Trauma, Medical conditions, Triggers, Trigger zones, Activity, pharmacologic

Evaluation and treatment history
Physicians and other health care providers

Psychosocial history
Substance use, Occupational and personal use, Sleep history
Clinical presentation of some conditions of secondary Headache:

Vascular Causes
Sub arachnoid hemorrhage [SAH] presents with
Severe HA of acute onset, Neck stiffness without fever
Supple neck does not exclude SAH

Presentations of SAH
- Headache present in 90% of cases
- Classic HA, sudden severe and continuous, often with nausea, vomiting, meningsmismus, focal neurological findings and loss of consciousness.
- Explosive HA, < worst HA of my Life >. Feeling of burst in 12%. Mild gradually increasing HA in 8%, sudden severe in 92%, can occur in any location, unilateral or bilateral.
- 33% present with HA only, 75% present with HA, nausea and vomiting.
- 66% sudden severe HA with loss of consciousness or focal deficits. 50% present with none or minimal HA and slight nuchal rigidity or moderate to severe HA with no neurological deficits or a cranial palsy. Stiff neck present in 75% cases during first 24 hrs and on fourth day after. 50% have presentation similar to meningitis, HA, stiffness of neck, nausea, vomiting, photophobia and low grade fever.
- Transient loss of consciousness in up to 33%

SENTINAL HA OR WARNING LEAKS:
This type occurs in 50% of cases before a major rupture of a saccular aneurysm, presents usually with unexplained sudden onset at any location. Duration is of 1 – 15 days.
50% only seek medical attention and they are misdiagnosed.

Temporal arteritis HA: Age of onset is over 50 yrs, mean age is around 70 yrs.

Presents as a sharp, dull, throbbing, burning lancinating pain, involving temples in 54% of cases. Associated with Polymyalgia Rheumatica, fever and weight loss and rapidly developing blindness. Normal ESR does not exclude the disease. It should be borne in mind that migraine is more prevalent in elderly than Giant Cell Arteritis.

Headache due to Stroke:
Among ischemic, CVA,-- Cerebro–venous thrombosis is commonly associated with HA. It is 80% in 213 cases of CVT. The thrombotic obstruction of Superior Sagittal Sinus elevates the intra cranial pressure by increasing the intravenous CSF pressure giving rise to HA and papilloedema. It is diffuse, progressive and constant.

HEADACHE DUE TO HYPERTENSION[HTN]:
Mild to moderate HTN usually does not cause HA. Severe HTN when diastolic>120mmHg can cause bi-occipital throbbing HA. It is often present in the morning on awakening. HTN due to following causes can cause HA....

92 % of Pheochromocytoma are associated with severe bilateral throbbing HA, malignant HTN, Acute Pressor response to exogenous agents like MAO inhibitors, red wine, pseudo ephedrine and toxemias of pregnancy.

ANGINAL HEADACHE:
Rare cause of HA due to cardiac ischemia noted on exertion and relief on rest. Unilateral or bilateral HA with or without accompanying chest pain. Mechanism is obscure. May be cardiac afferents joining tractus salitorious converging with cranio vascular afferents.

RAISED CSF PRESSURE HEADACHE:
Neoplasm is an uncommon cause of HA . Pituitary macro adenomas can present with trigeminal neuralgia, cluster like HA and RAEDEERS syndrome. Pituitary hemorrhage can be clinically silent or presents as migraine.
Persistent raised intracranial pressure can trigger chronic migraine. It is generalised HA present on waking improves as the day goes and worse with recumbency.

PSEUDOTUMOUR CEREBRI OR BENIGN INTRACRANIAL HTN:
Clinically Neurological Examination is normal except for the presence of papilloedema, visual loss and VI th nerve palsy. An elevated pressure with a normal CSF points by exclusion to diagnosis of PTC. The mechanism is postulated as due to increased rate of CSF formation and decreased rate of CSF absorption. The treatment is by ACETAZOLAMIDE a Carbonic Anyhydrase inhibitor which reduces CSF production. The clinical presentation is worst ever Bi-frontal pain which is a common location and orbital pain.

Features of idiopathic PTC
90% of pts are young obese woman. HA present in 75% or more. Papilloedema in 95 %. Cranial nerve VI palsy in 25%. Transient visual obscuration in 70%. Visual loss in 33%

LOW CSF-FLUID-HA:
Lumbar puncture is a common cause. Spontaneous occurrence also is well recognized. Infrequent causes are severe dehydration, DM coma, Uraemia, severe systemic infections. Besides LP index events include vigorous valsalva maneuver, multiple orgasms,
straining, coughing, cleaning the eustachian in Aeroplanes. It is a Bifrontal occipital throbbing HA occurring in a upright position decreasing or resolving when supine. Aggravated by cough, sneeze, straining and jugular vein compression. HA can be immediate or delayed for 14 days. Usually remits spontaneously within 5 days and in 20% may prolong up to 12 months. Mechanism not entirely known. It is explained that leakage through puncture exceeds the formation of CSF. It is low volume CSF than low pressure CSF.

**Paranasal sinus infections**

<table>
<thead>
<tr>
<th>Para nasal sinus</th>
<th>Possible locations of pain</th>
<th>Position which improves pain</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maxillary</td>
<td>Gums, maxillary teeth, peri orbital, supra orbital, temporal</td>
<td>Lying supine</td>
</tr>
<tr>
<td>Frontal</td>
<td>Periorbital, retro orbital, temporal</td>
<td>Head upright</td>
</tr>
<tr>
<td>Ethmoid</td>
<td>temporal, inner canthal area, midline behind the nose</td>
<td>Head upright</td>
</tr>
<tr>
<td>Sphenoid</td>
<td>Retro-orbital, frontal, temporal, vertex</td>
<td>Head upright</td>
</tr>
</tbody>
</table>

**METABOLIC DISORDERS HA:**

In Diabetes Mellitus, sometimes HA may be the presenting symptom due to the deranged glucose metabolism, occasionally in Diabetic KetoAcidosis, HA is a dominating presenting complaint. Hypothyroidism, 14% of cases present with HA. Anaemia presents with HA due to hyperdynamic circulation and cerebral anoxia. FEVERS of non cephalic origin can give rise to HA due to increased blood flow. Typhoid often presents with suboccipital HA. Chronic Malaria --- HA is not uncommon presentation. Pyelonephritis presenting with a severe HA is quite common.

Ha --- In acute Hypoxia with PaO2< 70mm within 24 hrs or chronic hypoxia persistently below that level can cause HA. High Altitude HA is due to decreased partial pressure of O2.

**PRIMARY HEADACHES:**

The diagnosis of PH is made after elimination of all the causes of Secondary HA by history and relevant investigations. By definition, PH are disorders in which HA and associated features occur in the absence of any exogenous cause.

**CLASSIFICATION OF PH**

1. Migraine, 2. Trigeminal Autonomic Cephalgia [TAC], 3. Tension type, 4. Other PHs.

Migraine is a form of sensory processing disturbance of brain stem, hypothalamus, and thalamus involving sensory sensitivity. Patient complaints of throbbing pain, but there is no reliable relation shown between vessel diameter and the pain. A good story ruined by the facts. It is no more vascular HA.

The key features are

- a. it is largely inherited
- b. Abnormal perception of otherwise normal circumstances such as light and sound.
- c. the attack is stereotypical when severe. Migraine Aura is defined as a total neurological disturbance manifested as visual, sensory or motor symptoms. Seen in 30% of pts and neurally driven.

**International Headache Society features of migrain**

Repeated episodic headache (4 to 72 hours) with the following features:

Any two of:

- Unilateral throbbing
- Worsened by movement
- Moderate or severe

Any one of:

- Nausea/vomiting
- Photophobia and phonophobia

**Vestibular Migraine:**

Recurrent attacks of vertigo that are caused by Migraine. HA is often absent. Presents with positional or non-positional vertigo accompanied by migraine symptoms. treatment is directed at migraine. 1/3 of pts of vertigo are Migraineurs.

Tension Type HA:

TTHA is of three forms. a) infrequent TTHA --- fewer than 12 HA days in a yr.

b. Frequent episodic TTHA --- 12—180 HA days in a yr.
c) Chronic TTHA --- at least 180HA days per yr. The pain is bilateral in 90% of cases. It is pressing, band like discomfort, typically builds slowly, fluctuates in severity without accompanying features such as nausea, vomiting, photo and phonophobia. If at all one or two features may be associated in mild form. It is featureless and less disabling than Migraine.

The pathophysiology is pericranial myofacial pain sensitivity is increased and nociception is an important factor in TTHA. Conversion of episodic TTHA to chronic TTHA is due to sensitization of central nociceptive arteries and hence the role of Amitryptaline in treatment. Basically TTHA is disorder of pain modulation.

**TRIGEMINAL AUTONOMIC CEPHALGIA (TAC):**

Group of primary headache syndromes, all marked by headache and associated autonomic features. They are cluster headache, paroxysmal hemicrania, hemicrania continua, short lasting unilateral neuralgiform headache attacks with conjunctival injection and tearing syndrome – SUNCT. Diagnosis is clinical, basing on headache frequency, duration and accompanying symptoms. The brainstem connection between the trigeminal system and the parasympathetic autonomic nervous system allows us to understand how the trigeminal based pain can be associated with autonomic symptomatology.

Cluster Headache: It is 0.1% in population. Stereotype episodic severe, never mild, always unilateral head pain located retro orbitally in 70% cases and peri orbital and occipito-nuchal in
the rest. The quality is boring, stabbing, burning or squeezing associated with autonomic features such as lacrimation (31%) eyelid ptosis, nasal rhinorrhoea (42%) conjunctival injection (60%) and Horner’s syndrome. Phenomenon of unilateral photo phobia / phono phobia is characteristic of TAC. The duration is 15 Minutes to 180 Minutes usually less than 60 minutes. Peak time occurrence is 1 AM to 2 AM, 1 PM to 3 PM, after 9.00 PM- 60 to 90 minutes after sleep showing predilection to rapid eye movement phase. Cluster period or cluster cycle is time during which patient is having daily cluster headache. Usually the patient experiences 1 or 2 cluster periods in year. The number of attacks are One attack a week to 8 or more per day. The cluster period can last from 2 days to 2 weeks. During the period of attack patient is in a state of agitation because remaining still makes the pain worse. The patient crawls, bangs head, takes hot showers, develops suicidal ideation. Patient has his own setup during attack, and presses on eye with a hand. There is a clock like regularity in timing of attacks suggestive of dysfunction of hypothalamus, biologic clock mechanism. Periodicity is core feature of cluster headache. Next attack recurs about the same hour each day for the duration of cluster bout. Pain free interval is usually one year. Cluster headache is likely to be disorder-involving central pain maker neurons in the region of posterior hypothalamus.

INDOMETHACIN – RESPONSIVE TRIGEMINAL AUTONOMIC CEPHALGIAS:

Paroxysmal hemicrania presents with frequent unilateral severe boring clawlike high frequency orbital, temporal pain, all over the head radiating to neck and ipsilateral shoulder, lasting for 2 to 30 minutes with no predilection for nocturnal attacks, associated with lacrimation (62%) conjunctival injection (36%) rhinorrhoea triggered by rotating the head. Hemicrania continua – only 90 cases reported in the literature. It is continuous daily headache for 24 hours / day and 7 days in a week.

Short lasting unilateral nuralgiform headache attacks with conjunctival injection and tearing syndromes. It is very rare, many headache specialist have not seen this. Attacks of moderate to severe pain with conjunctival injection, tearing, nasal obstruction, localize to orbital or peri orbital areas. Mean age of on set is 51 years. Usually unilateral, average duration of pain is 10 to 60 seconds.

Other primary headaches: The pathogenesis is poorly understood. Some time they may be due to symptomatic structural lesions. The primary stabbing headache is shortest duration. The frequency is 1 to 50 times a day. The duration is 1 to 2 seconds. No trigger factors, no autonomic symptoms. The primary cough headache occurrence is 1% in general population. The posterior fossa tumors, platybaisa, basilar impressions, arnold–chiari manifestation have to be excluded. Primary exertional headache is brought by prolonged exercise. It is pounding, throbbing headache with nausea, vomiting. Photo and Phono Phobia. Pheochromocytoma, cardiac ischemia to be excluded.

Primary sexual headache: more common in men. There are three types a) Dull bilateral headache intense at orgasm b) sudden severe explosive headache occurring at orgasm c) postural headache developing after coitus. 5-12% SAH are precipitated by sexual intercourse. Hypnic headache, a sleep related primary headache developing after coitus. 5-12 % SAH are precipitated by sexual intercourse. Hypnic headache, a sleep related primary headache developing after coitus.

Clinical Features of the Trigeminal Autonomic Cephalgias

<table>
<thead>
<tr>
<th>Gender</th>
<th>M&gt;F</th>
<th>F=M</th>
<th>F~M</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pain</td>
<td>Stabbing, boring</td>
<td>Throbbing, burning</td>
<td>Burning, stabbing</td>
</tr>
<tr>
<td>Type</td>
<td>Excruciating</td>
<td>Excruciating</td>
<td>Severe to excruciating</td>
</tr>
<tr>
<td>Severity</td>
<td>Orbit, temple</td>
<td>Orbit, temple</td>
<td>Periorbital</td>
</tr>
<tr>
<td>Site</td>
<td>1/alternate day</td>
<td>1-40/d (&gt;5/d for more than half the time)</td>
<td>3-200/d</td>
</tr>
<tr>
<td>Attack frequency</td>
<td>B/d</td>
<td>15-180 min</td>
<td>2-30 min</td>
</tr>
<tr>
<td>Duration of attack</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes (prominent conjunctival injection and lacrimation)</td>
</tr>
<tr>
<td>Autonomic features</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Migrainous features</td>
<td>Yes</td>
<td>Yes</td>
<td>Yes</td>
</tr>
<tr>
<td>Alcohol trigger</td>
<td>Yes</td>
<td>No</td>
<td>No</td>
</tr>
<tr>
<td>Cutaneous triggers</td>
<td>No</td>
<td>No</td>
<td>Yes</td>
</tr>
<tr>
<td>Indomethacin effect</td>
<td>-</td>
<td>Yes</td>
<td>-</td>
</tr>
<tr>
<td>Abortive treatment</td>
<td>Sumatriptan injection or nasal spray Oxygen</td>
<td>No effective treatment</td>
<td>Lidocaine (IV)</td>
</tr>
<tr>
<td>Prophylactic treatment</td>
<td>Verapamil</td>
<td>Indomethacin</td>
<td>Lamotrigine</td>
</tr>
<tr>
<td>Treatment</td>
<td>Methysergide</td>
<td>Topiramate</td>
<td>Gabapentin</td>
</tr>
</tbody>
</table>

Classification of Chronic Daily Headache

<table>
<thead>
<tr>
<th>&gt;4 H Daily</th>
<th>&lt;4 H Daily</th>
<th>Secondary</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chronic migraine</td>
<td>Chronic cluster headache</td>
<td>Posttraumatic</td>
</tr>
</tbody>
</table>

731
### Headache as a Symptom

Headache as a symptom is seen as manifestation of almost every system disorder of the body. Headaches also can be there without any structural disorder otherwise grouped as primary headaches. It is obvious that once the neuro imaging study is normal, history can alone help at determining origin of headache. The international headache society classification described the criteria for diagnosis of various types of headache, basing on clinical history. The diagnosis can be arrived like mathematical calculation if the laid down criteria is applied. With the availability of neuro imaging facility at every major taluk or mandal, management of headache can be more rational, exciting, enthusiastic and beneficial to the patient, if the physician familiarizes well with all the types of headache and the associated features.

### REFERENCES: