Special Headache

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Outline of Special Headache

- I. Other primary Headache
  - I-4.3. Promary Exertional Headache
  - I- 4.5. Hypnotic Headache
- II The Secondary Headache
  - II- 6.7.8. Headache attributed to RCVS
  - II- 7. Headache attributed to Non- Vascular Intracranial Disorder
    - II-7.3.3 Antiphospholipid syndrome (APS)
  - II- 10. Headache attributed to disorder on homeostasis
    - II-10.1.1 High –Altitude Headache
  - II- 11. Cranium-facial Headache
    - II-11.2.1 Cervicogenic Headache
- III. Cranial Neuralgia/Facial pain
  - III-13.8. Occipital Neuralgia

I. Other primary Headache

I-4.3 Primary Exertional Headache

- Exertional Headache: 1. Promary EH 2.secondaryEH
- ICHD-II Criteria:

  Description: Headache precipitated by any form of exercise
- Diagnosis Criteria:

  A. Pulsating headache fulfilling criteria B&C

  B. Lastigng from 5 minutes to 48hrs

  C. Brought on by ocurring only during or after physical exertion

  D. Not attributed to another disorder

Pathophysiology of primary EH

- Unknown

- Vascular origin (hypothesis)

  Arterial dissection

  Venous distension

  Internal jugular vein valve incompetence

D.D

- Secondary Headache

  Subarachnoid hemorrhage (SAH)

  Posterior fossa lesion: Chiari malformation type 1, tumor

  Cardiac cephalgia: Coronary ischemia, angina pectoris

  Spontaneous intracranial hypotension (SIH)

  Venous sinus stenosis or occlusion

Work up and Treatment (Exertional Headcahe)

- Neuroimaging recommended with:

  Cough HA/ Exertional HA/HA associated sexual activity (Taiwan Headache Society Guideline)

- Cardiac cephalgia survey if Dx of exertional HA in older or cardiovascular risk factor p'ts
- Treat primary EH:

  With self-limiting course

  Most adolescents do not use pain killer (Taiwan)

  Behavior modification : Avoid/reducing precipitatiing factors

  When exrtional is unavoidable: Indomethcin (no control trial)

I- 4.5. Hypnotic Headache (H.H)

Relationship Between Sleep and Headache

- **Sleep-related headaches (during or after sleep)**
- **Sleep-phase–related headaches**
  - **III, IV, REM: migraine**
  - **REM: cluster headache, chronic paroxysmal hemicrania**
- **Length of sleep and headaches: migraine**
  - **Excessive deep sleep**
  - **Lack of sleep**
  - **Sleep disruption**

Relationship Between Sleep and HA-2

- **Sleep relieves headaches**
  - Migraine and other types of headaches
- **Sleep disorders and headaches**
  - Sleep apnea and headaches
  - Somnambulism and headaches
  - Other parasomnias and headaches
- **Effect of headaches on sleep**
  - Minimal to major sleep disruption
- **Dreams and headaches**

Relationship between HA and Sleep

1. Headache is the result of disrupted nocturnal sleep or the underlying process that disrupts sleep. (sleep disturbance is the cause of HA)
   a. Obstructive sleep apnea (OSA), UARS or nocturnal hypoxia or hypercarbia
   b. Restless legs syndrome or periodic leg movements of sleep (PLMS)
   c. Psychophysiologic insomnia
   d. Chronic pain syndrome or fibromyalgia
   e. Depression or anxiety

Relationship between HA and sleep

2. Headache is the "cause" of a disturbance of nocturnal sleep.

   a. Chronic tension-type headache (more than 15 headache days per month for more than 6 months)

   b. Chronic migraine with or without analgesic abuse or depression or anxiety

Hypnic Headche (I-4.5)(睡眠頭痛)

- 診斷基診
- A.　頭痛為鈍痛，符合基準B.D
- B.　只有在睡眠中產生，並使病人醒來
- C.　至少具下列兩項特徵:

  每個月內發作>15 次　醒來後持續>15分鍾

  首次發作在50 歲之後
- D.　無自主神經系統的症狀且下列症狀最多不超過一項:噁心、畏光或怕吵
- E.　非歸因於其它疾患
- 应排除顱內疾患. 為有效處理病人,辨別該頭痛與三叉自律神經痛Trigeminal autonomic cephalgias是必要的

Clinical comparison of major TACs

	Cluster headache	Paroxysmal hemicrania	SUNCT syndrome
Epidemiology			
Gender (male:female)	3:1	1:3	8:1
Prevalence	0.9%	0.02%	Very rare
Age of onset	28–30 years	20–40 years	20–50 years
Pain			
Quality	Boring, throbbing	Boring	Stabbing
Intensity	Extremely high	High	Moderate to high
Localization	Periorbital	Orbital, temporal	Orbital, temporal
Duration of attack	15–120 min	2–45 min	5–250 s
Frequency of attack	1–8/day	1–40/day	1/day to 30/h
Autonomic symptoms	+ +	+ +	+
Circadian rhythmicity	+	(–)	-
Alcohol trigger	+ +	(+)	(–)

SUNCT, short-lasting unilateral neuralgiform pain with conjunctival injection and tearing. Modified from Ref. [2]. - = none; (–) = rare; (+) = infrequent; + = modest; + + = strong.

II. The secondary HA

II- 6.7.8. Reversible Cerebral Vasoconstriction Syndromes(RCVS)

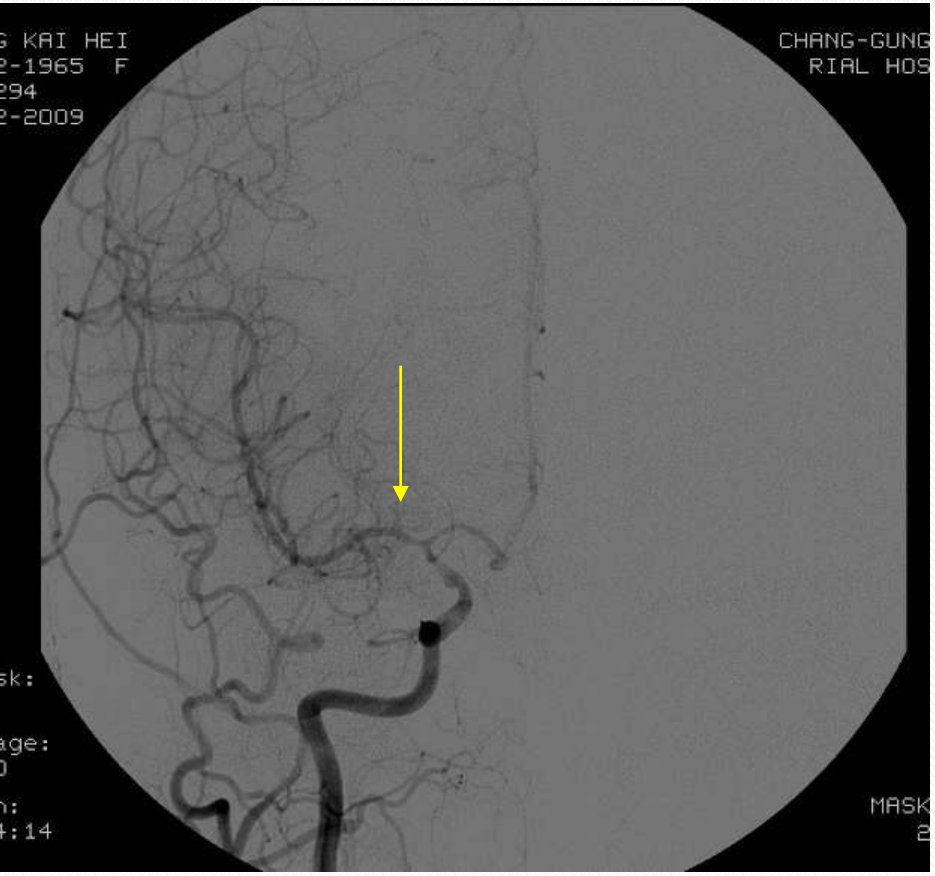
- Severe headaches with or without seizures and focal neurological deficits
- A group of disorders
  - Thunderclap headache with vasospasm
  - Benign angiopathy of the central nervous system (BACNS)
  - Migrainous vasospasm or crash migraine
  - Call-Fleming syndrome (or Call syndrome)
  - Postpartum angiopathy
  - Drug-induced cerebral vasoconstriction

Reversible Cerebral Vasoconstriction Syndromes (RCVS)

- Female predominance (around 80 %)
- Middle-aged:40~50 year-old
  - Female older than male patients
- Blood pressure surges (SBP>160mmHg) in 1/3 patients
- Headache duration: 1-3 hours
- Disease course: 1-3 months
- Risk factors:
  - History of migraine
  - In the postpartum period
  - Exposed to different pharmacologic agents

Thunderclap headache(6.4)

- Diagnostic criteria:
  - (A) Severe head pain fulfilling criteria B and C
  - (B) Both of the following characteristics:
    - Sudden onset, reaching maximum intensity < 1 min
    - Lasting from 1 h to 10 days
  - (C) Does not recur regularly over subsequent weeks or months
- Associated diseases: intracranial aneurysm, cerebral venous thrombosis, cervical artery dissection, spontaneous intracranial hypotension, pituitary apoplexy, reversible cerebral vasoconstriction syndromes…….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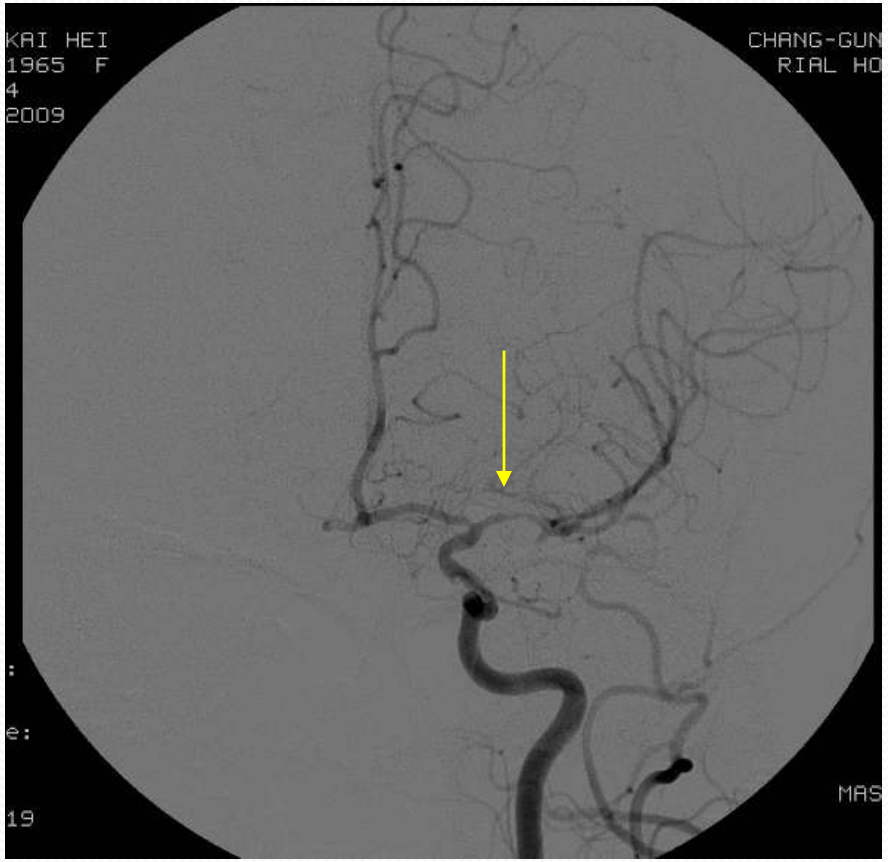
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Etiologies of RCVS-(1)

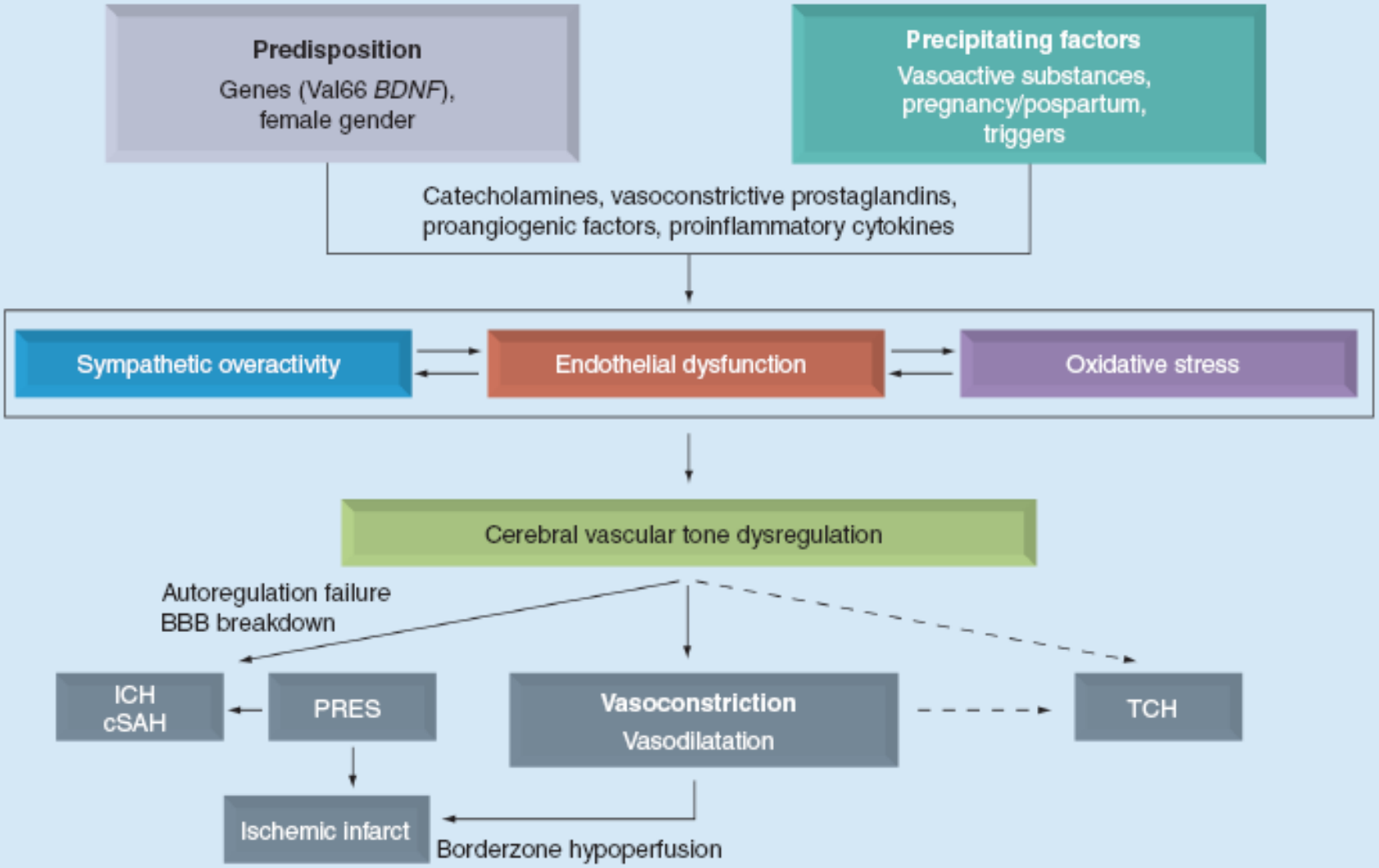
- *Primary*
  - Spontaneous or evoked by triggers
    - Primary thunderclap headache
    - Exertional headache
    - Headache associated with sexual activity
    - Cough headache
    - Bath-related thunderclap headache

Etiologies of RCVS-(2)

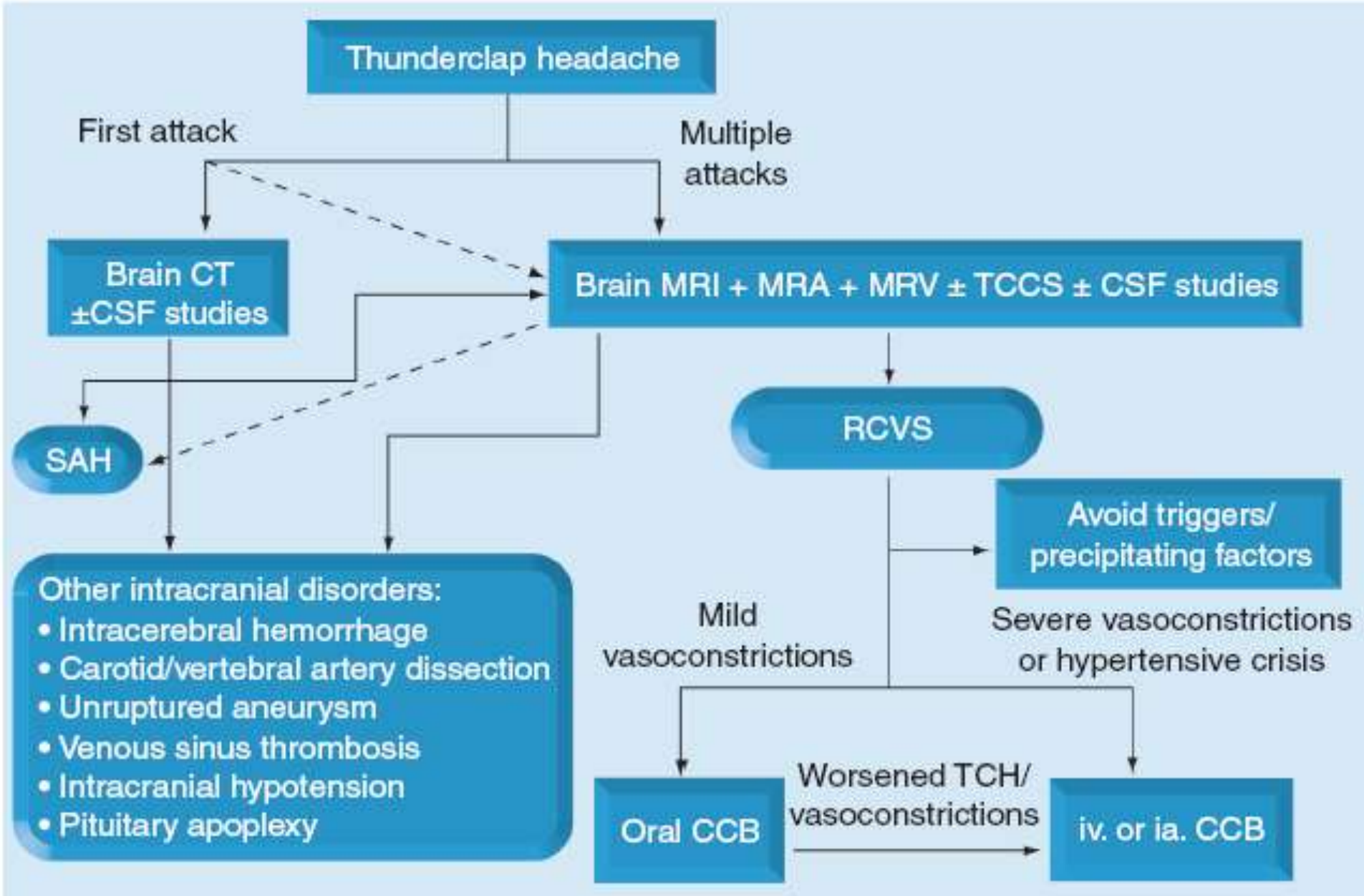
- *Secondary*
  - Vasoactive substances
    - Illicit drugs: ***Cannabis***, cocaine, ecstasy, amphetamines, LSD
    - Ergots: ***Ergotamine***, methergine, methylergometrine, lisuride, bromocriptine
    - Sympathomimetics:Ephedrine, isometheptene, ***pseudoephedrine***, diet pills, phenylpropanolamin
    - Serotonergic drugs: ***Selective serotonin-reuptake inhibitors(SSRI), triptans***
    - Immunosuppressants:Tacrolimus (FK-506), cyclophosphamide, IFN-a
    - Others: Nicotine patches, ***Ginseng***, licorice, indomethacin, binge drinking, oral contraceptive pills, hormonal ovarian stimulation for intrauterine insemination

Etiologies of RCVS-(3)

- *Secondary*
  - Pregnancy and the postpartum period
  - Catecholamine secreting tumors
  - Extra- or intra-cranial arterial disorders or procedures
  - Blood products
  - Intracranial disorders or surgery
  - Miscellaneous :Hypercalcemia, systemic lupus erythematosus
  - Genetic study: Brain-derived neurotrophic factor (BDNF)



Expert Rev. Neurother. 11(9), 1265–1276 (20



Expert Rev. Neurother. 11(9), 1265–1276 (20

II. The Secondary HA

II.7.3.3 Headache attributed to Non- Vascular Intracranial Disorder

Antiphospholipid syndrome (APS)

Abstract

The antiphospholipid syndrome is an autoimmune disease characterised by recurrent arterial or venous thrombosis, pregnancy morbidity and the persistence of positive antiphospholipid antibodies. Many other clinical manifestations may occur including heart valve disease, livedo reticularis, thrombocytopenia and neurological manifestations such as migraine and seizures. We review a number of other manfestations including stenotic lesions, coronary artery disease and accelerated atherosclerosis, skeletal disorders and the concept of seronegative antiphospholipid syndrome.

Table 1. Classification of patients with antiphospholipid syndrome according to the underlying condition*

Underlying condition	No. (%) of patients
Primary antiphospholipid syndrome	531 (53.1)
Systemic lupus erythematosus	362 (36.2)
Lupus-like syndrome	50 (5.0)
Primary Sjögren's syndrome	22 (2.2)
Rheumatoid arthritis	18 (1.8)
Systemic sclerosis	7 (0.7)
Systemic vasculitis	7 (0.7)
Dermatomyositis	5 (0.5)

* There was an overlap of 2 conditions in 2 patients.

Table 2. Clinical features at disease onset in 1,000 patients with antiphospholipid syndrome*

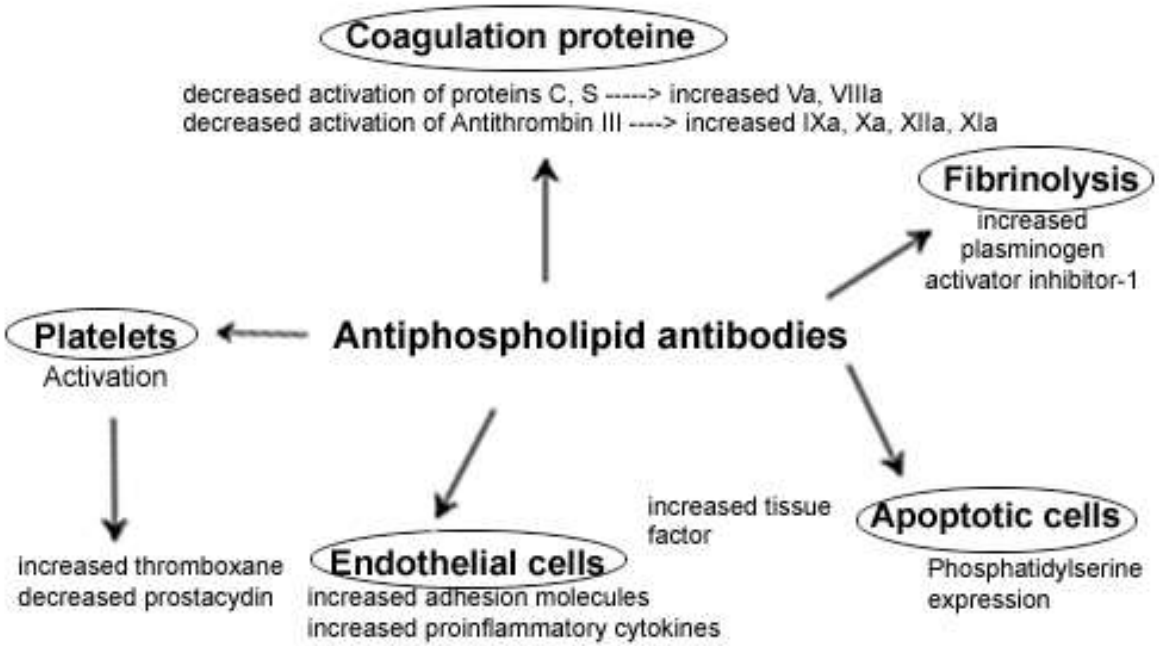
Manifestation	No. (%) of patients
Deep vein thrombosis	317 (31.7)
Thrombocytopenia (<100,000 platelets/μl)	219 (21.9)
Livedo reticularis	204 (20.4)
Stroke	131 (13.1)
Superficial thrombophlebitis	91 (9.1)
Pulmonary embolism	90 (9.0)
Fetal loss	83 (8.3)
Transient ischemic attack	70 (7.0)
Hemolytic anemia	66 (6.6)
Skin ulcers	39 (3.9)
Epilepsy	34 (3.4)
Pseudovasculitic skin lesions	26 (2.6)
Myocardial infarction	28 (2.8)
Amaurosis fugax	28 (2.8)
Digital gangrene	19 (1.9)

* Some patients had several associated presenting manifestations.

Table 4. Immunologic findings in 1,000 patients with antiphospholipid syndrome

Parameter	No. (%) of patients
Anticardiolipin antibodies	879 (87.9)
IgG and IgM	321 (32.1)
IgG alone	436 (43.6)
IgM alone	122 (12.2)
Lupus anticoagulant	536 (53.6)
Alone	121 (12.1)
With anticardiolipin antibodies	415 (41.5)
Antinuclear antibodies	597 (59.7)
Anti–double-stranded DNA	292 (29.2)
Anti-Ro/SSA	140 (14.0)
Anti-La/SSB	57 (5.7)
Anti-RNP	59 (5.9)
Anti-Sm	55 (5.5)
Rheumatoid factor	78 (7.8)
Cryoglobulins	36 (3.6)

Figure 1. Pathogenetic mechanims of antiphospholipid antibodies
M. Tektonidou, et al. QJM 2000.9:523-30



APS: Diagnostic Criteria

Requires at least one clinical and one laboratory criteria

- **Clinical Criteria**

1) Vascular thrombosis

At least one episode of arterial, venous, or small-vessel thrombosis in any tissue or organ Confirmed by imaging or biopsy

2) Pregnancy morbidity (≥1 of the following)

At least one unexplained death of morphologically normal fetus ≥10 weeks gestation

At least one premature birth (<34 weeks) of morphologically normal neonate due to eclampsia, severe pre-eclampsia, or placental insufficiency

At least three consecutive spontaneous miscarriages prior to 10 weeks gestation, otherwise unexplained

J Thromb Haemost2006;4:295-306

APS: Diagnostic Criteria

Laboratory Criteria

- Positive lupus anticoagulant on at least two occasions, at least 12 weeks apart **(+)**
- Medium or high titer IgG or IgM anti-cardiolipin antibody on at least two occasions, at least 12 weeks apart
- Medium or high titer IgG or IgM anti-B2 glycoprotein I antibody on at least two occasions, at least 12 weeks apart

J Thromb Haemost2006;4:295-306

“Non-Criteria” Manifestations

- ***Neurologic***
- ***Cardiac***
- Pulmonary: Diffuse alveolar hemorrhage
- Hematologic: Thrombocytopenia, Hemolytic anemia
- Dermatologic: Livedo reticularis, Raynaud phenomenon

Lupus2010;19

II.The secondary HA

II-10.1.1 Headache attributed to disorder on homeostasis

High –Altitude Headache

- ICHD-II: 10.1.1. The headache occurs within 24hrs after acute onset of hypoxia with PaO2 less than 70mmHg or in chronically hypoxic p'ts withPa02 persistently at or below this level
- Headache attributed to a disorder of homeostasis: Hypoxia, Hypercapnia, High Altitude Headache, Diving (altitude decompression sickness), Sleep apnea HA, Dialysis, Arterial H/T, Pheochromocytoma, Hypertensive crisis without encephalopathy, Hypertensive encephalopathy. Pre-eclampsis, Eclampsisa, Hypothyroidism, Fasting, Cardiac cephalgia, Other
- C/F of HAH

A: >-2/5of following and fulfilling criteria C/D: 1)Bilateral 2) F or F/T 3) Dull or moderate intensity 4) Aggravated by exertion, movement, straining, coughing or bending

B: Ascent to altitude >2500m

C: Headache develops within 24 hrs after ascent

D: Headache resolves within 8 hrs after descent

High Altitude Headache

Clinical Presentation

Altitude illness is divided into three syndromes:

- Acute mountain sickness (AMS)
- High-altitude cerebral edema (HACE)
- High-altitude pulmonary edema (HAPE)
- **Pre-Existing Medical Problems** be advised to consult a physician familiar with high-altitude medical issues

High Altitude Headache

Box 2-3. Tips for acclimatization

- Ascend gradually, if possible. Try not to go directly from low altitude to >9,000 ft (2,750 m) sleeping altitude in one day.
- Consider using acetazolamide (Diamox) to speed acclimatization if abrupt ascent is unavoidable.Avoid alcohol for the first 48 hours.
- Participate in only mild exercise for the first 48 hours.
- Treat an altitude headache with simple analgesics.
- Know the early symptoms of altitude illness: Headache is the cardinal symptom, sometimes accompanied by fatigue, loss of appetite, nausea
- Dexamethasone and pulmonary artery pressure-lowering drugs, such as nifedipine or sildenafil(Viagra), may be carried for emergencies.

1I.The secondary HA

1.2: Headache attributed to disorder of neck

II.11.2.1: Cervicogenic headcahe (CH)

A. Pain from cervical, refer to head and or face

B. From clinic, laboratory, image revealed headache from cervical spine or cervical soft tissue change

C. Revealed the pain due to nuchal disease (any one):
1. Pain form neck in clinic sign. 2. Under control: Pain relieved by diagnostic nerve block

D. Pain relieved within 3 months after treat the pathogenic disease

5.3/4:Acute/Chronic headache attributed to whiplash injury

- 5.3:Acute/Chronic headache

  A. Headache, no known typical headache (include C and D)

  B. Whiplash with neck pain history

  C. Headache attack within 7 days after whiplash injury

  D. Headache remission within 3 months after whiplash injury

- 5.4: Chronic headache attributed to whiplash injury ICHD-II

  Headache persisted for more than 3 months after whiplash injury

Table 9-4. Sequelae of whiplash injuries

- Neck and back injuries
  - Myofascial
  - Fractures and dislocations
  - Disc herniation
  - Spinal cord compression
  - Spondylosis
  - Radiculopathy
  - Facet joint syndrome
  - Increased development of spondylosis
- Headaches
  - Tension type
  - Greater occipital neuralgia
  - Temporomandibular joint disorder
  - Migraine
  - Third occipital headache
- Dizziness
  - Vestibular dysfunction
  - Brain stem dysfunction
  - Cervical origin
  - Barré syndrome
  - Hyperventilation syndrome
- Paresthesias
  - Trigger points
  - Thoracic outlet syndrome
  - Brachial plexus injury
  - Cervical radiculopathy
  - Facet joint syndrome

TABLE 9-4. *Continued*

- Carpal tunnel syndrome
- Ulnar neuropathy at the elbow
- Trigeminal sensory impairment

Weakness
- Radiculopathy
- Brachial plexopathy
- Entrapment neuropathy
- Reflex inhibition of muscle contraction by painful cutaneous stimulation

Cognitive, somatic, and psychological sequelae
- Memory, attention, and concentration impairment
- Nervousness and irritability
- Sleep disturbance
- Fatigability
- Depression
- Personality change
- Compensation neurosis

Visual symptoms
- Convergence insufficiency
- Oculomotor palsies
- Abnormalities of smooth pursuit and saccades
- Horner's syndrome
- Vitreous detachment

Rare sequelae
- Torticollis
- Tremor
- Transient global amnesia
- Esophageal perforation and descending mediastinitis
- Hypoglossal nerve palsy
- Superior laryngeal nerve paralysis
- Cervical epidural hematoma

Internal carotid and vertebral artery dissection

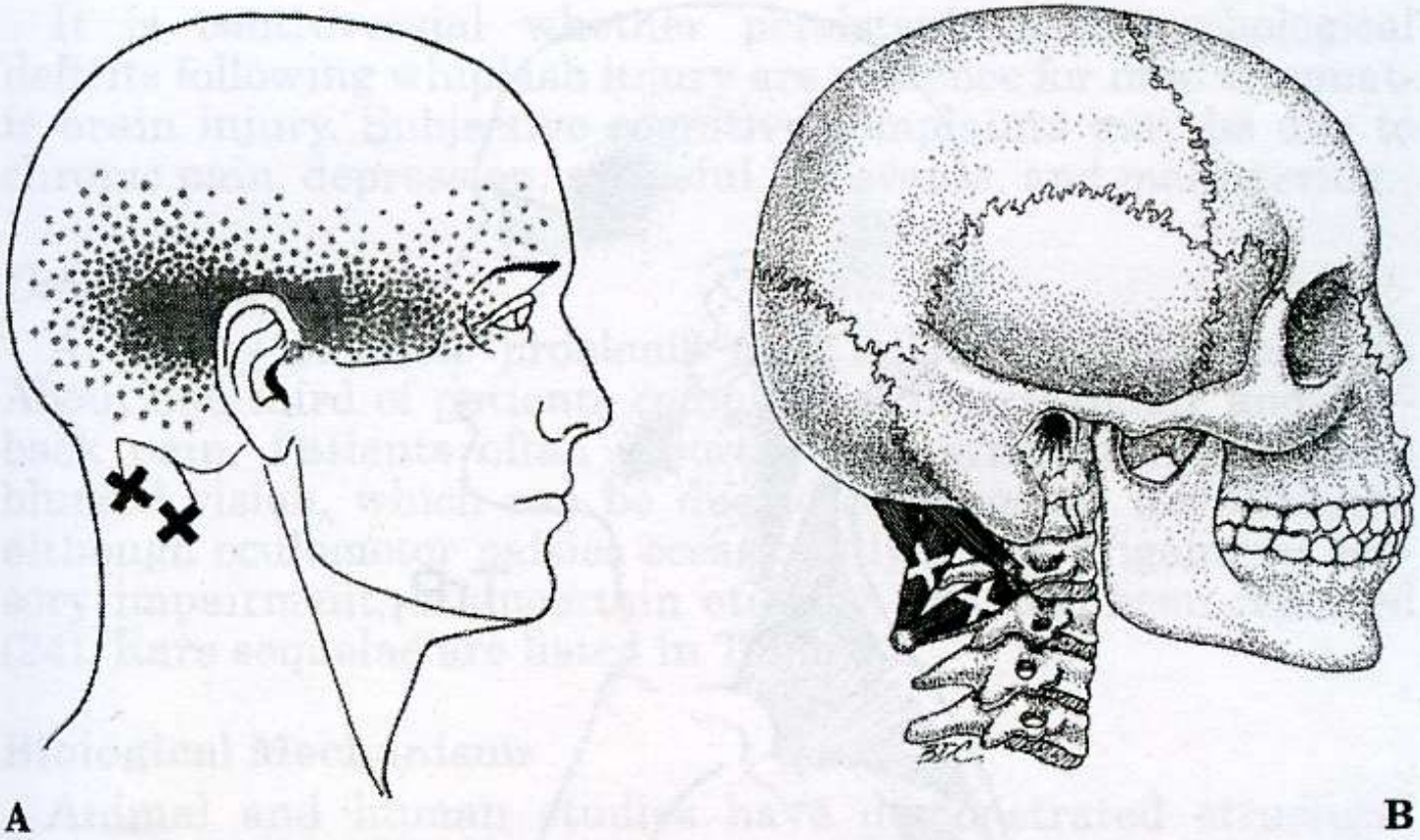


Figure 9-3. Referred pain pattern (A) of trigger points (Xs) in the right suboccipital muscles (B). (From Travell JG, Simons DG. *Myofascial pain and dysfunction: the trigger point manual.* Baltimore: Williams & Wilkins, 1983:322, with permission.)

Table 9-6. Percentages of patients with persistence of neck pain and headaches following a whiplash injury

Symptom	1 week	3 months	6 months	1 year	2 years
Neck pain	92	38	25	19	16
Headache	57	35	26	21	15

Cervicogenic headcahe

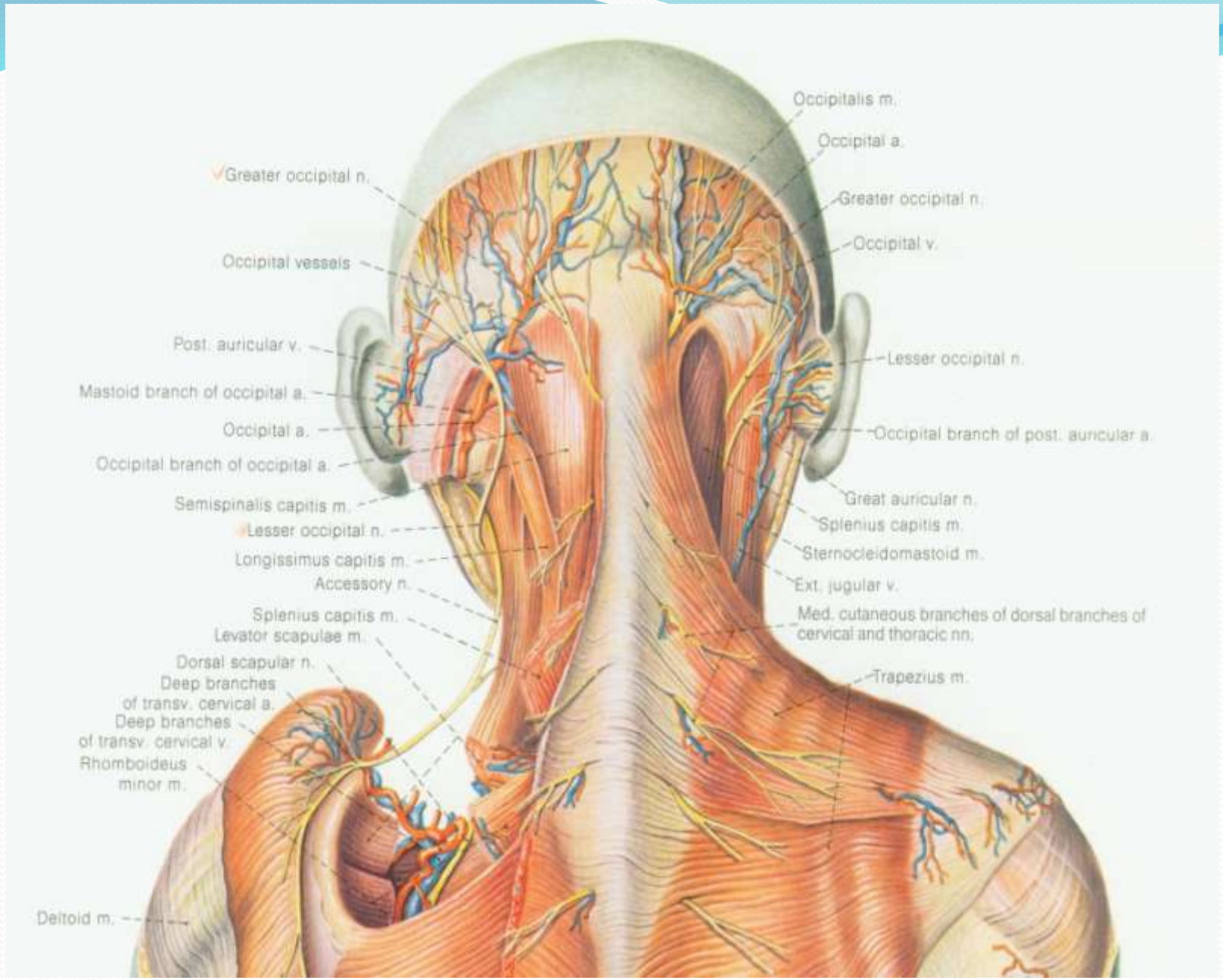
- Cervical spondylosis and muscle spasm may cause headache in older pt's. Unilateral or bilateral.
- Digital pressure in suboccipital area may produce the headache
- The headache may due to occipital neuralgia, MFS with triggor point and referred pain from neck structure such as upper cervical facets
- Tx: NSAIDs, M. relaxant, TCA, physical therapy. Occipital nerve block and triggor point injection may helpful in appropriate cases

III.13.8. Occipital Neuralgia (O.N)

- Misnomer? The pain isn't necessarily from occipital nerve and dosen't usually have a neuralgic quality
- Anatomy:

1) Greater occipital nerve (GON)- mainly dorsal ramus of C2, minor of C3, to medial part of scalp pierce fascia below superior nuchal ridge to vertex, along with occipital artery.

2) lesser occipital nerves (LON)- ventral primary rami of C2-3, along postreior border of SCM, divide to cutaneous brabch (lateral portion of scalp), and cranial surface of pinna of ear (From C2-3 cervical nerves)


Occipitalis m.
Occipital a.
Greater occipital n.
Greater occipital n.
Occipital v.
Occipital vessels
Post. auricular v.
Lesser occipital n.
Mastoid branch of occipital a.
Occipital a.
Occipital branch of post. auricular a.
Occipital branch of occipital a.
Great auricular n.
Semispinalis capitis m.
Splenius capitis m.
Lesser occipital n.
Sternocleidomastoid m.
Longissimus capitis m.
Ext. jugular v.
Accessory n.
Med. cutaneous branches of dorsal branches of cervical and thoracic nn.
Splenius capitis m.
Levator scapulae m.
Trapezius m.
Dorsal scapular n.
Deep branches of transv. cervical a.
Deep branches of transv. cervical v.
Rhomboideus minor m.
Deltoid m.

Occipital neuralgia (O.N)-3

Etiology of O.N

- 1. Most O.N are idiopathic
- 2. Related to other specific causes: Trauma, Prior skull base surgery, Rheumatoid arthritis, Nerve entrapment

Compression: i) Entrapment of the greater occipital nerve in the aponeurosis of superior trapezius or semispinalis capitis muscle. ii) Hypertrophied atlantoepistophic ligament (C1- 2). iii) By anomalous ectasic verteral artery. iv) Degenerative C1-2 arthrosis pathology- C2-3 facet joint, other upper cerviacal spine pathology, posterior fossa

2) Non compression: i) Referred from the trigger points (muscle spasm) in superior trapezius, semispinalis capitis or other suboccipital muscles. ii) O.N (pain on C2-3 region) evoked by V1-2 facial herpes zoster infection, respiratory infection, iii) Other headache: TTHA, Migraine, Cluster HA, other cranial pain iv) Cervical myelitis, multiple sclerosis

Diagnosis of Occipital Neuralgia

- ICH-II: 1) Paroxysmal stabbing pain on greater occipital nerve (G.O.N) and lesser O.N (L.O.N) 2) Tenderness on G/L.O.N

  3) Local anesthetics blockade make transient pain relief
- Neural blockade of greater and lesser occipital nerves : Diagnostic maneuver to help confirm of O.N and distinguish from Tension type headache (TTHA). Due to O.N easily be blocked at nuchal ridge, but not response to TTHA.
- D.D: Atlantoaxial joint and upper zygoapophysial joint or cervical muscles tenderness with refer pain

Treatment of Occipital Neuralgia (O.N)-1

- Neural blockade with local anesthetic and steroid combined with NSAIDS, muscle relaxants, TCA and physical therapy
- BTX injection: G.O.N and trigeminal neuralgia (T.N). – Cephalagia 2005 (case report)
- Refractory O.N: Preoperative assessment with CT-guided nerve block prior to dorsal cervical rhizotomy. (Better response in p’ts without prior head or neck surgery)- AJNR Am J Neuroradiol 2003

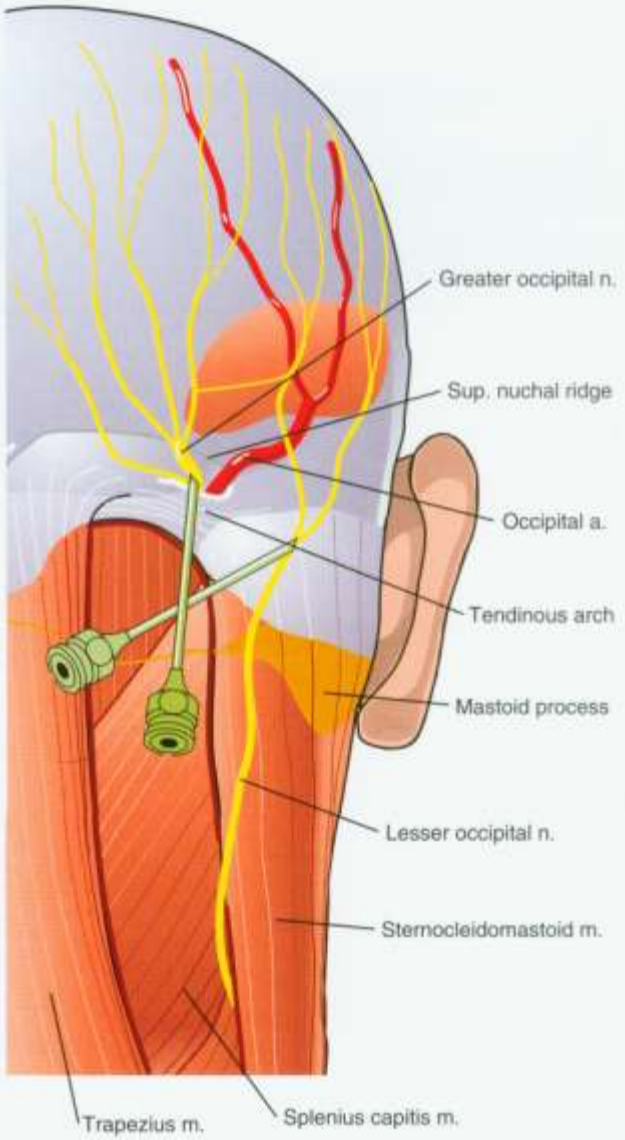


Figure 6–2. Proper needle placement for greater and lesser occipital nerve block. (From Waldman SD: Atlas of Interventional Pain Management. Philadelphia, WB Saunders, 1998, p 21.)