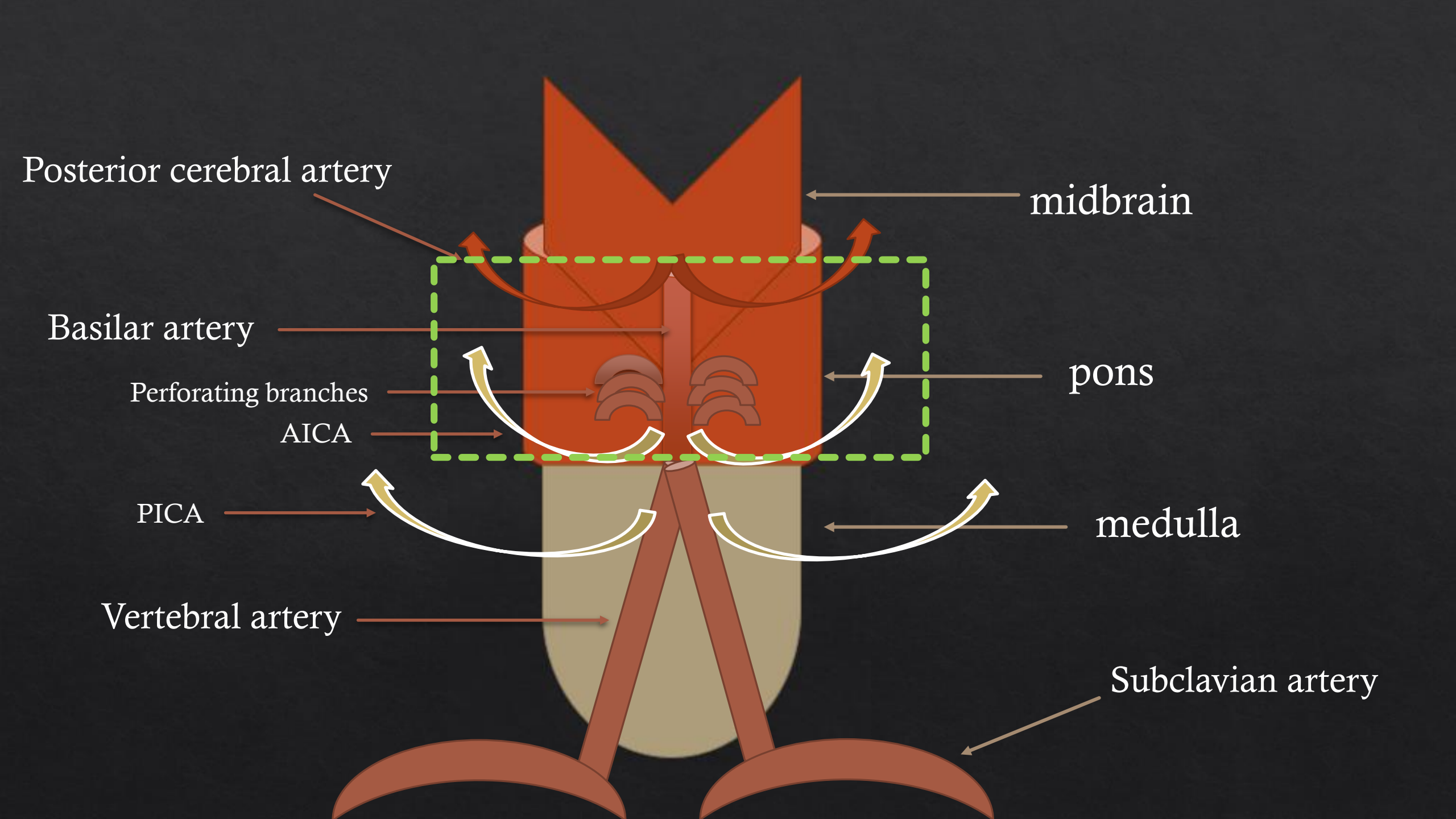
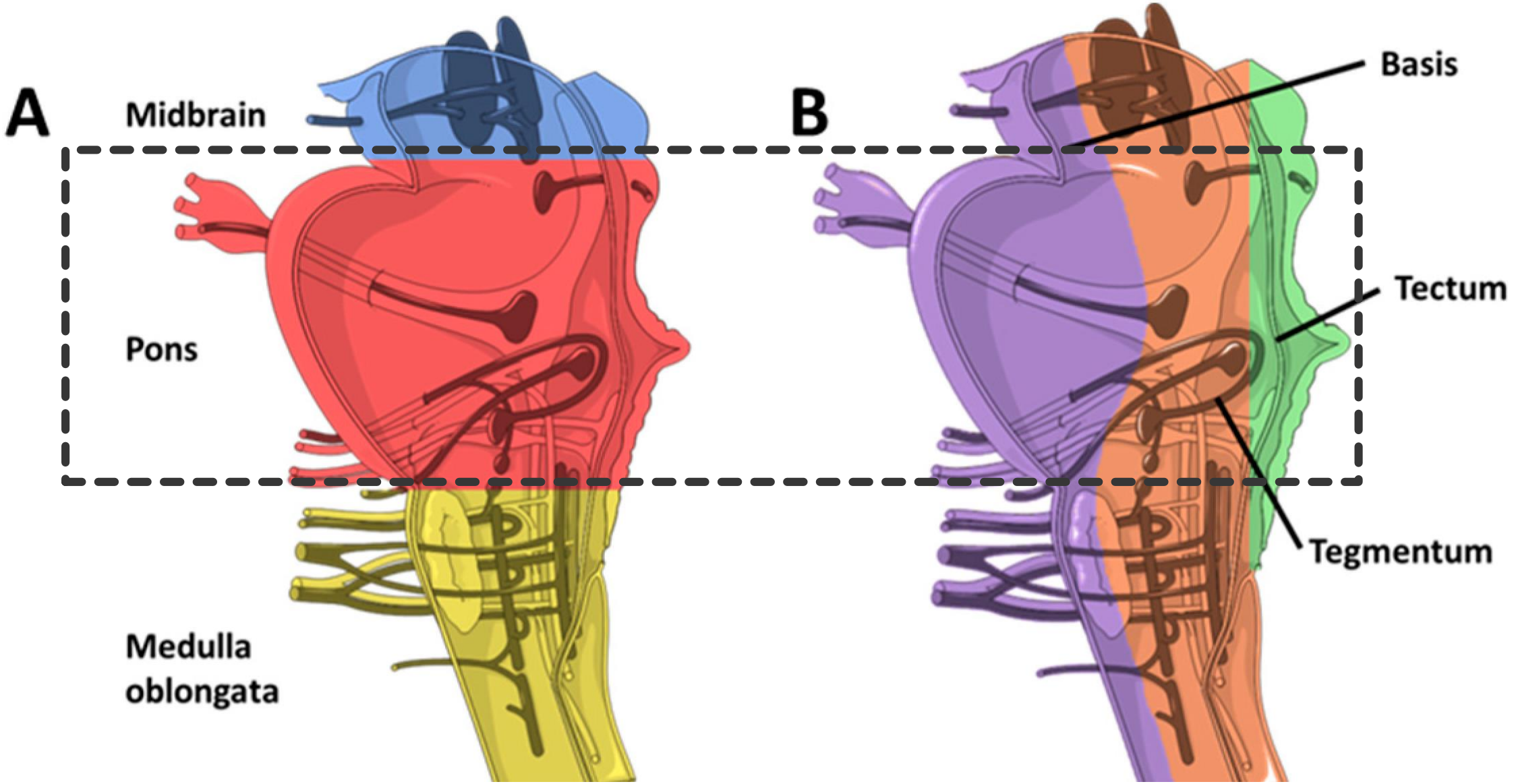
Locked-in-Syndrome (aka. pseudocoma)

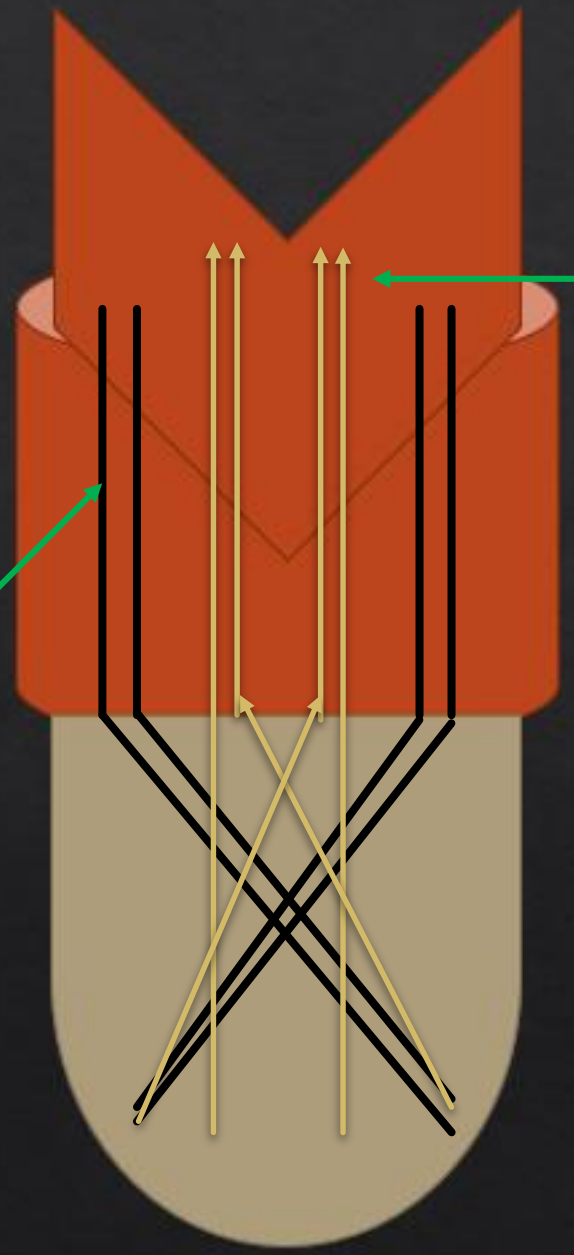
Munmun Aziz
Oct. 24. 2020


Posterior cerebral artery
midbrain
Basilar artery
pons
Perforating branches
AICA
PICA
medulla
Vertebral artery
Subclavian artery


A
Midbrain
Pons
Medulla
oblongata
B
Basis
Tectum
Tegmentum

Descending fibers [UMN]

- Corticospinal tracts
- Corticopontine
- Corticobulbar

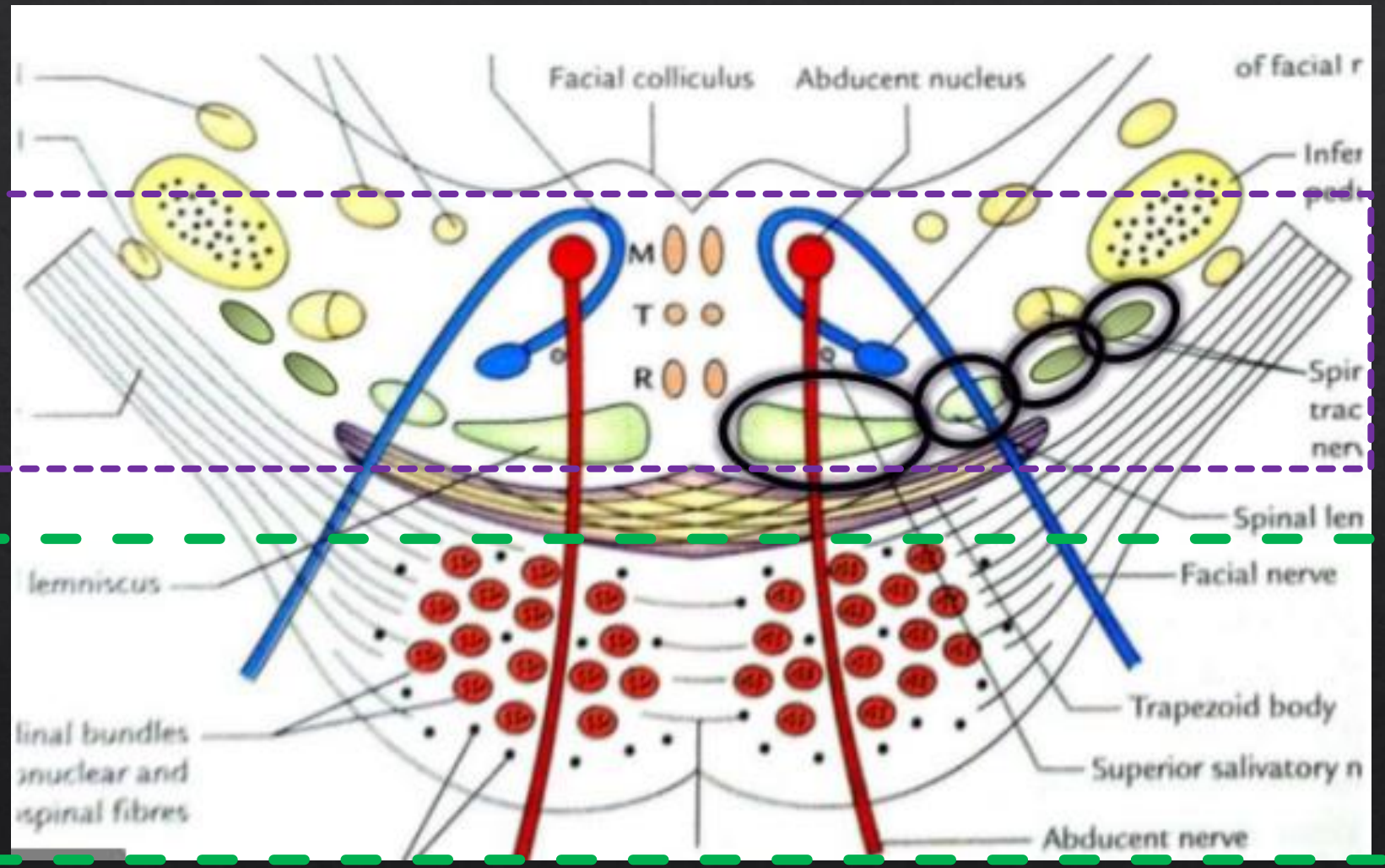


Ascending fibers [sensory]

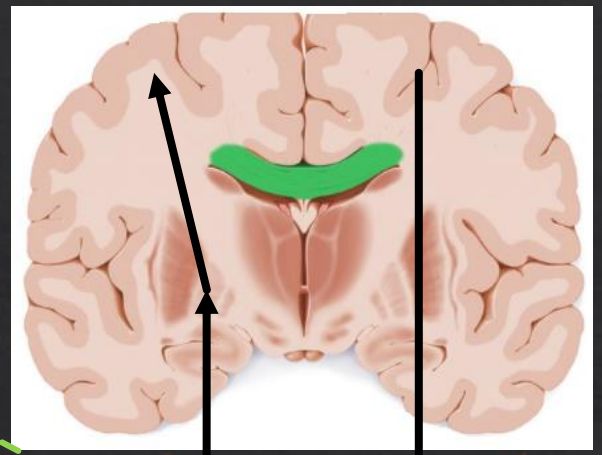
- Dorsal column fibers
  - Fine touch/2-point discrimination
  - Proprioception
  - Vibration
- Spinothalamic tract
  - Pain
  - temperature

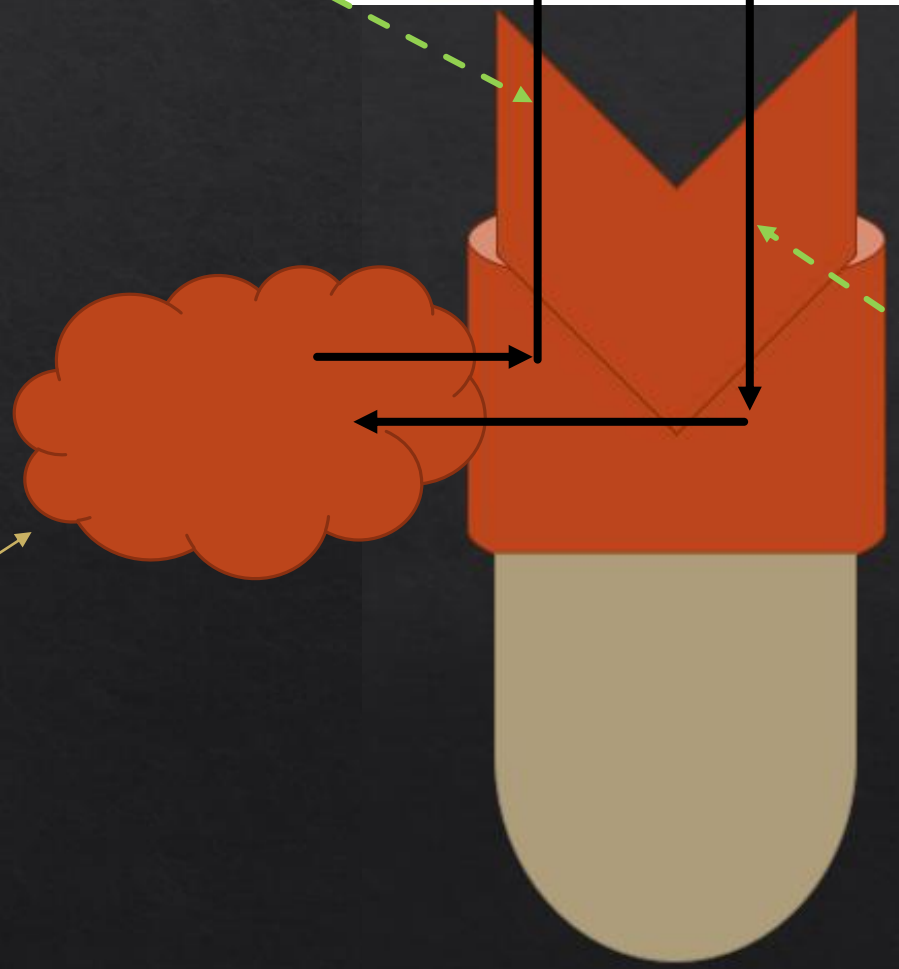
Ascending fibers
[Tegmentum]

Descending fibers
[basis]



Cerebello-thalamo-cortical tract





cerebellum

Cortico-ponto-cerebellar tract

Additional tracts that pass-through PONS

Vestibular nucleus [pons]

↓

Vestibulospinal tracts

↓

Extensor muscles

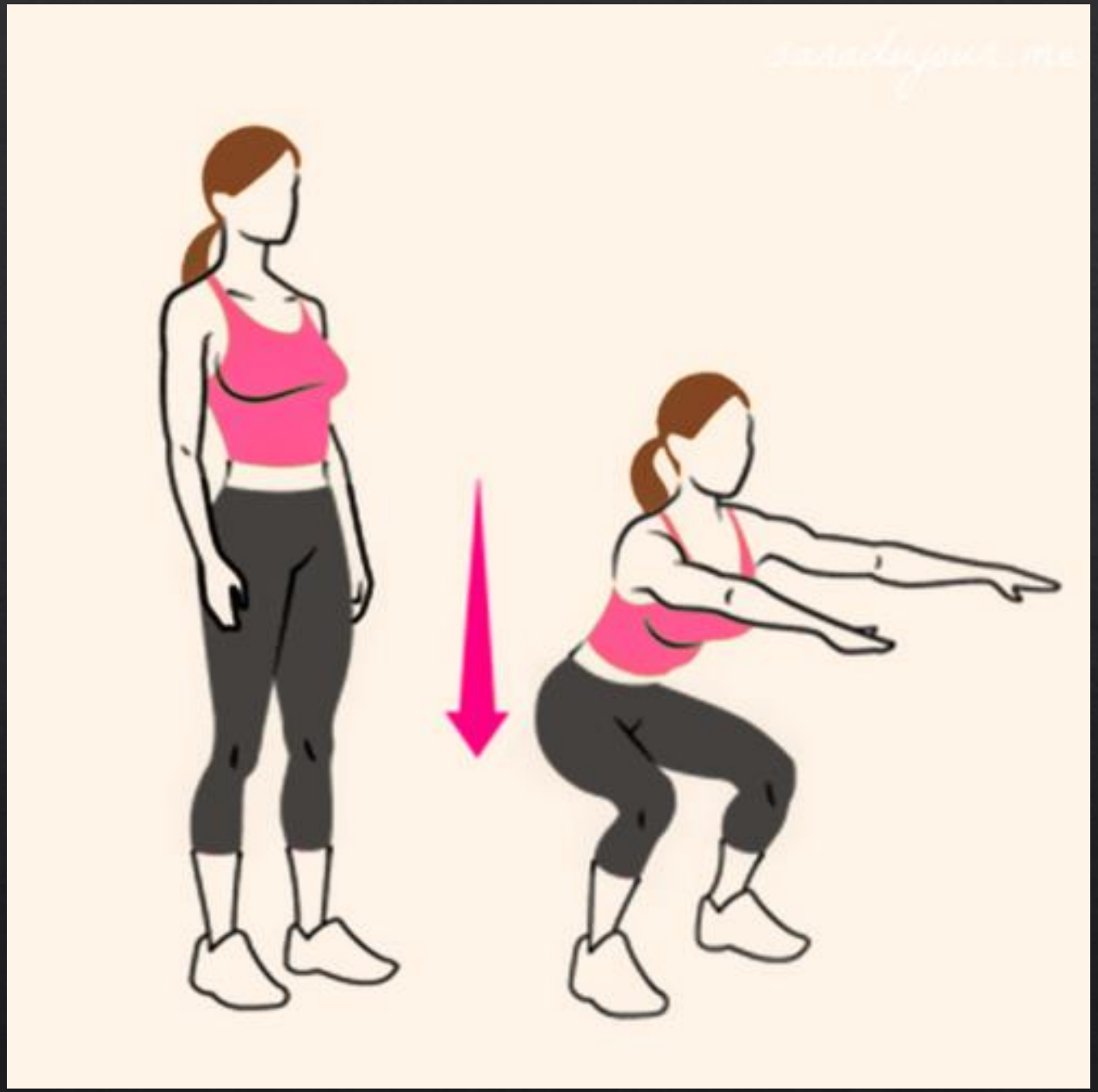
Red nucleus midbrain

↓

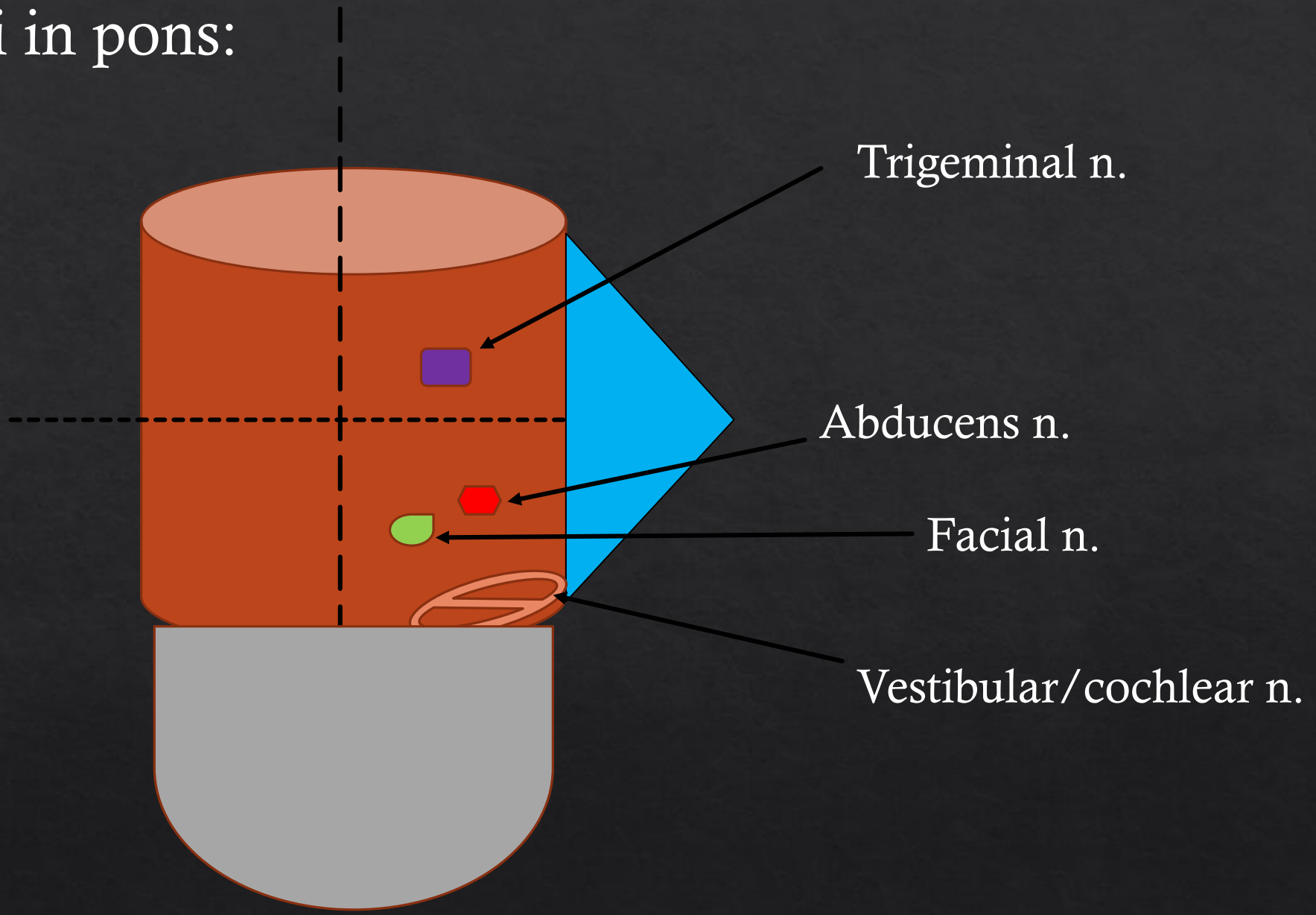
Rubrospinal tract

↓

Flexor muscles



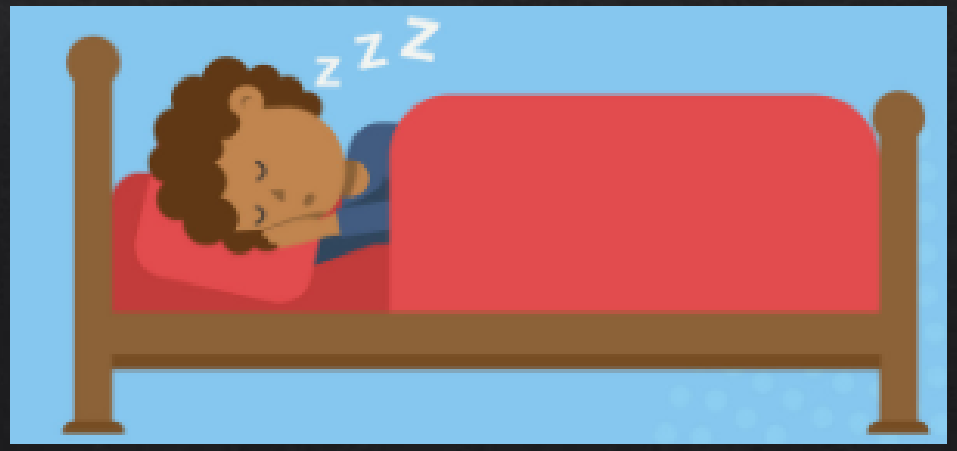
Cranial nerve nuclei in pons:



What is purpose of reticular formation?

- Allows you to alternate between slow sleep rhythms [NREM] and fast sleep rhythms [REM].

- Also responsible for arousal and wakefulness.

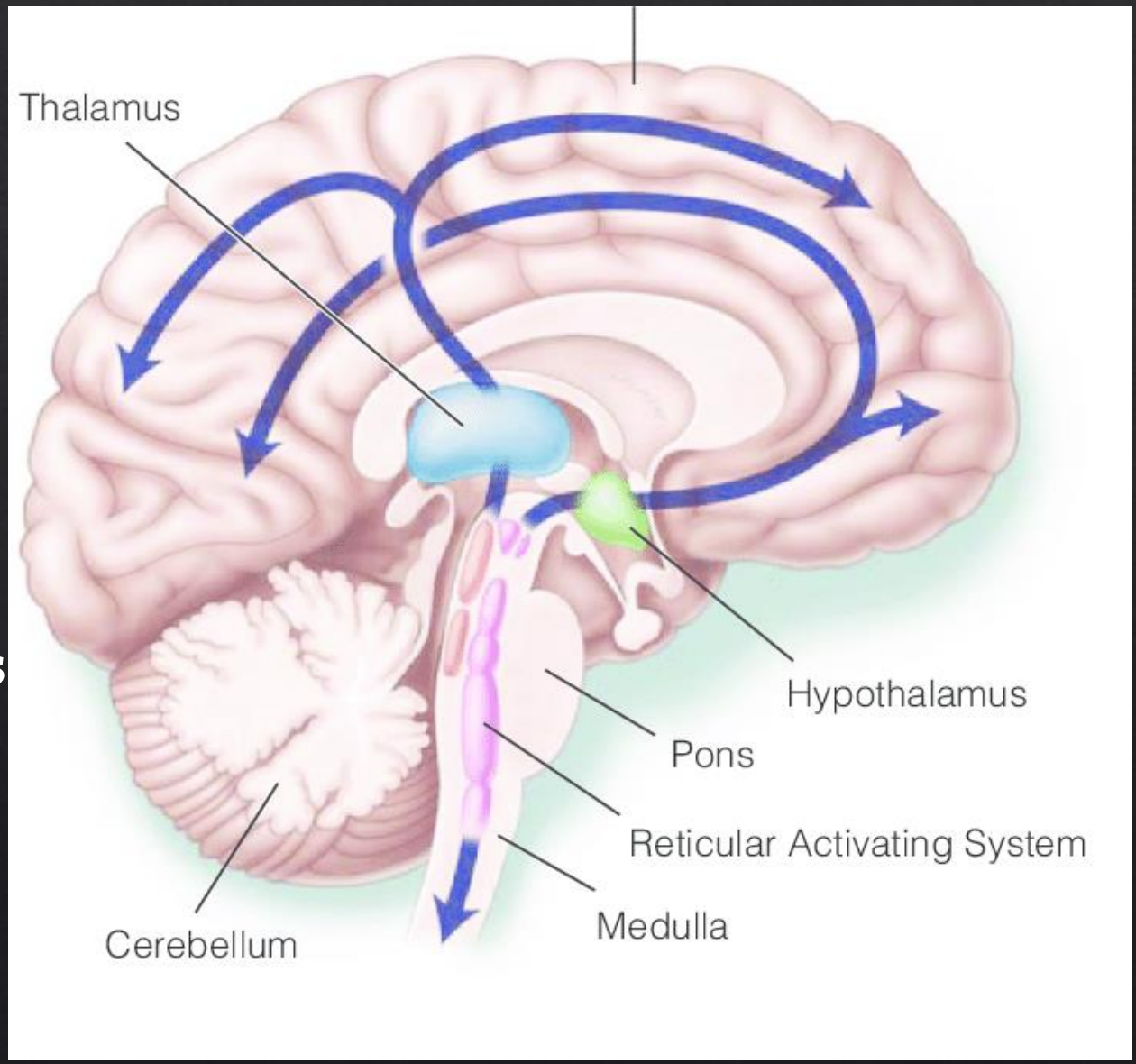


4 components of Reticular formation:

- **Locus coeruleus**
  - Location→ dorsolateral pons of the brainstem
  - Neurotransmitter: NE
- **Raphe nuclei**
  - Locations→ midline throughout brainstem
  - Neurotransmitter: serotonin
- **Posterior tuberomammillary hypothalamus**
  - Location: Posterior aspect of brainstem
  - Neurotransmitter: histamine
- **Pedunculopontine tegmentum**

  Location: Midbrain & pons

  Neurotransmitter: cholinergic



What is Locked-in syndrome[LIS]?

LIS occurs due to a lesion affecting the ventral pons (less commonly the midbrain), resulting in…

Classic form:

- Quadriplegia
  - Corticospinal tract impaired
- Bulbar palsy (speech and swallowing deficit)
  - Cortico-nuclear fibers impaired
- Whole body sensory loss

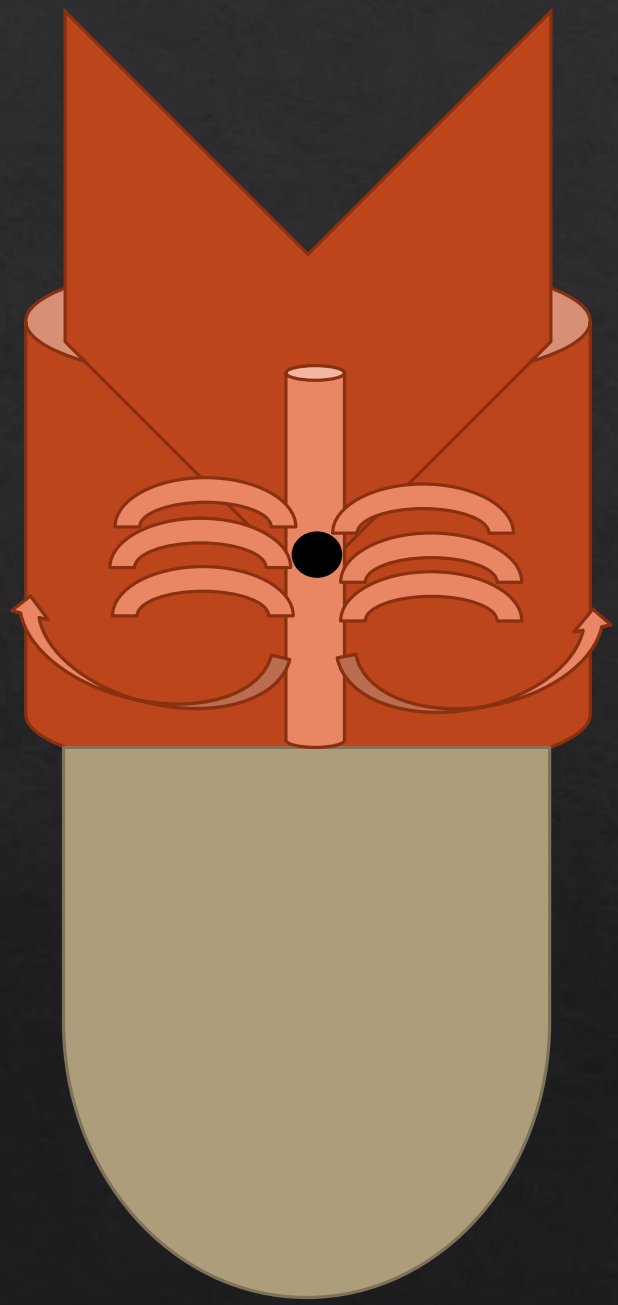
Etiology:

- Vascular (most common cause in older patients)
  - Ischemic stroke (ex. basilary artery occlusion; hypotension)
  - Hemorrhagic stroke

- Trauma (2nd common; see in children)
  - Blunt/Penetrating

Less common causes:

- Mass (ex. metastasis from adenocarcinoma of lung)
- Infection (ex. pseudomonas predomin. pontine abscess)
- Demyelination (ex. central pontine myelinosis)



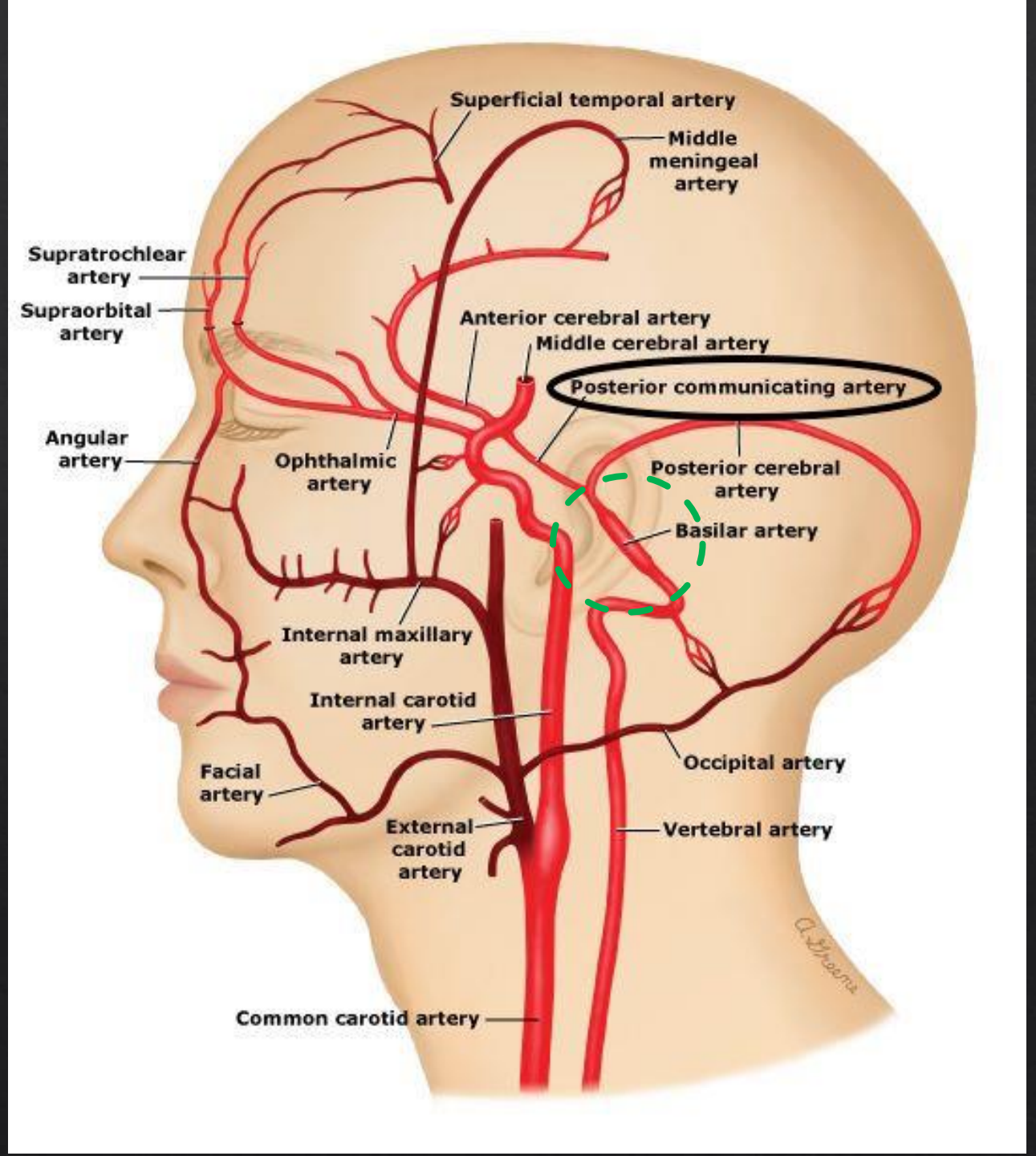
What is preserved in LIS?

Classic form:

- Cognition/conscious
- Vertical eye movement/upper eyelid movement/blinking
- Hearing

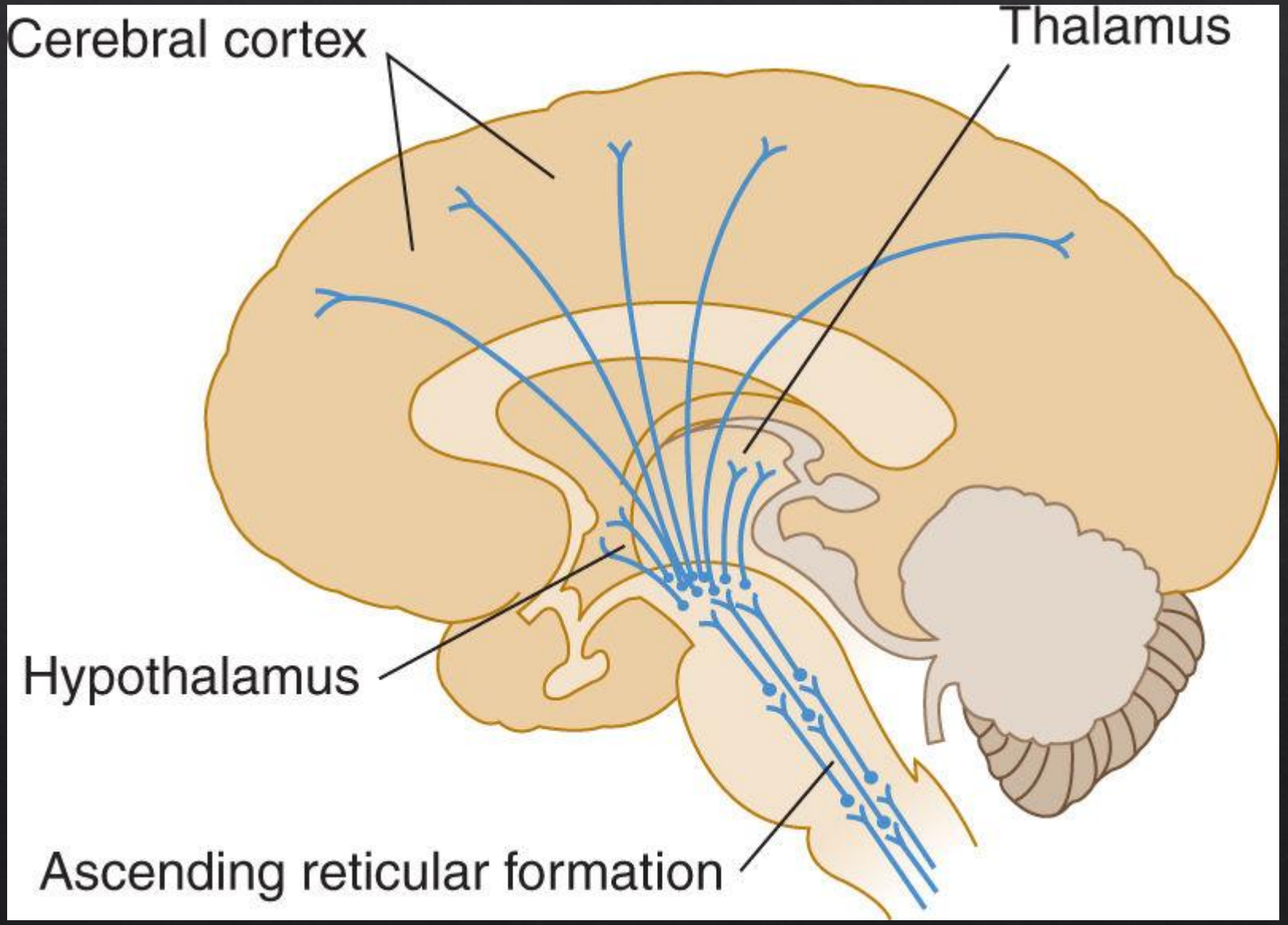
How is cognition intact?

Anterior cerebral circulation & posterior cerebral circulation connected via posterior communicating artery



How is patient with LIS conscious?

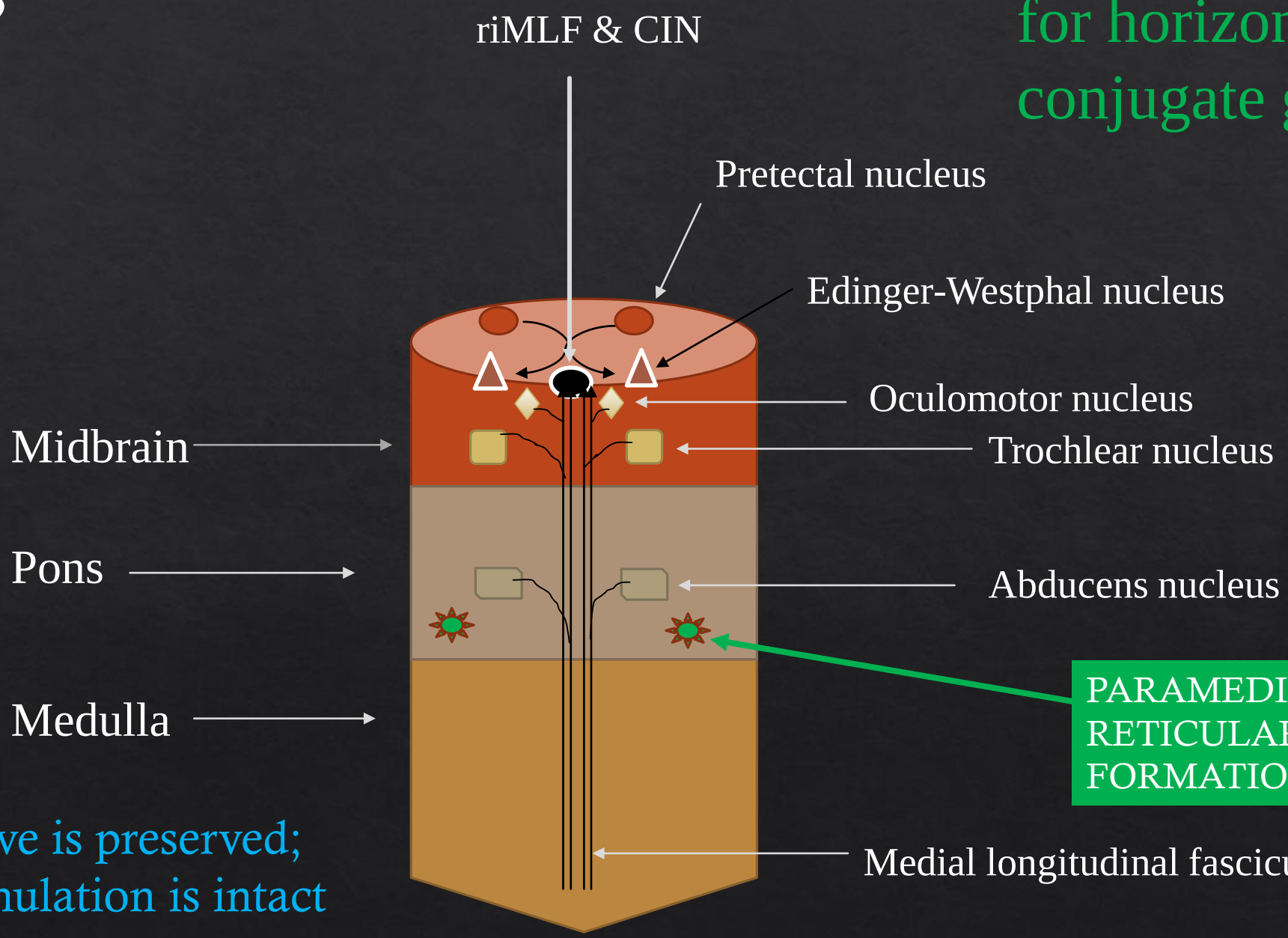
Reticular formation is functioning.



How is vertical gaze intact?

midbrain

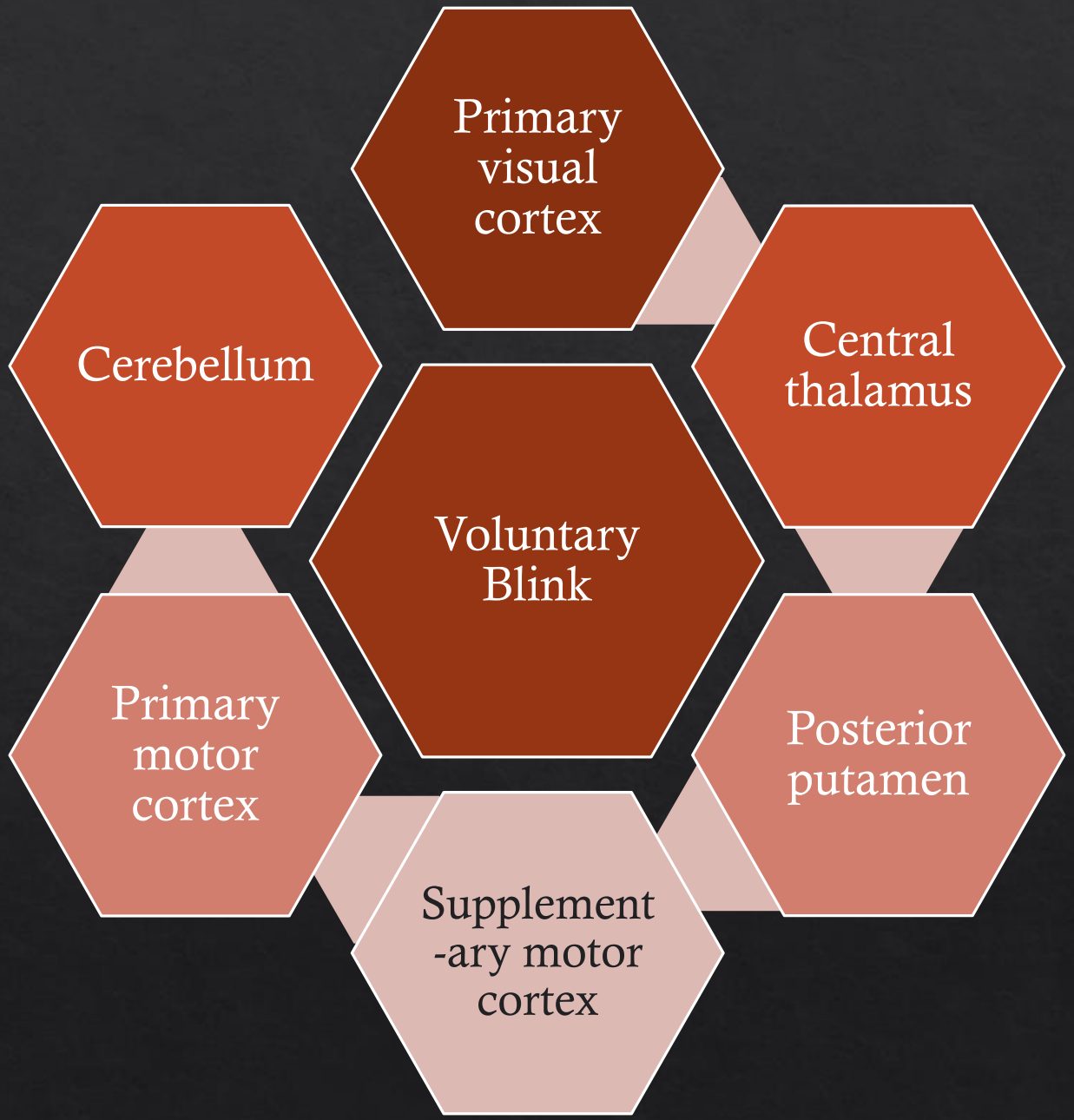
What is responsible for horizontal conjugate gaze?



Oculomotor nerve is preserved; upper eyelid stimulation is intact

How is blinking intact?

- Voluntary* (done consciously)
- Reflex (involves pons; ex. Corneal reflex, auditory reflex)
- Spontaneous (normal; preventing cornea from drying)

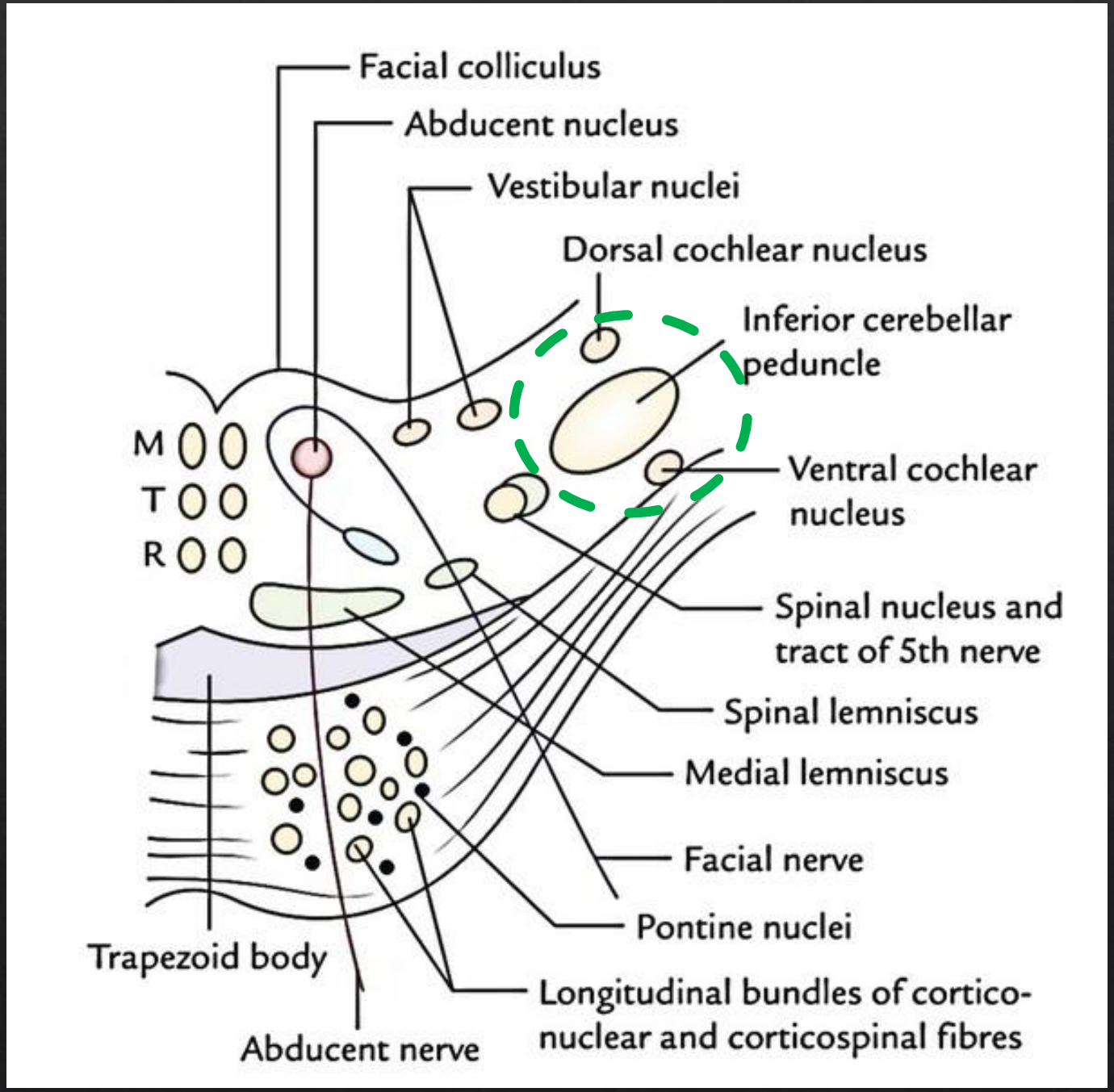


(Smit, 2008)

How is hearing intact?

The pontomedullary junction is supplied by the confluence of:

- Vertebral a
- Basilar a.
- PICA



(Mercier et. al, 2008)

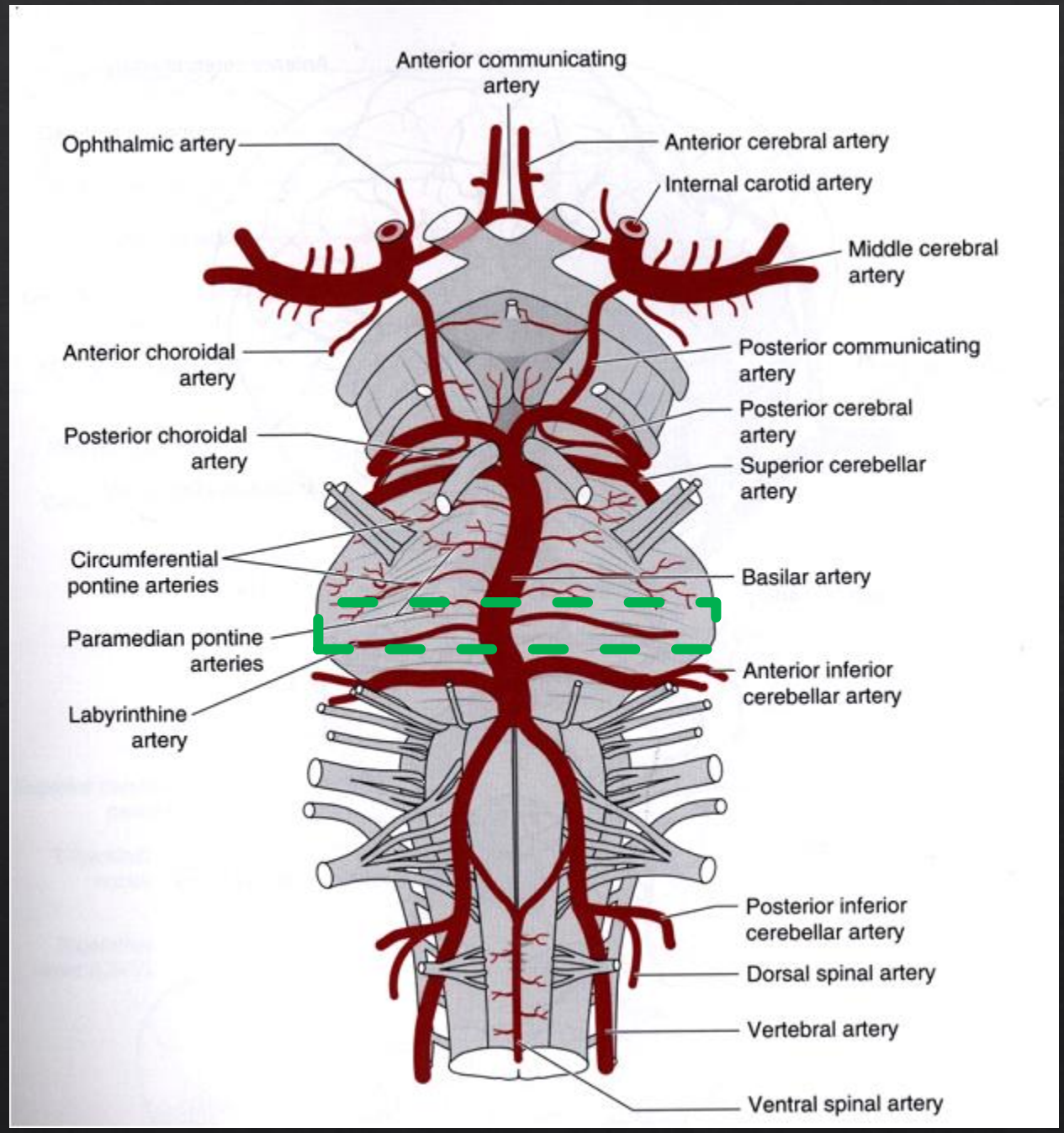
Inner ear blood supply:

- Labyrinthine artery (branch of the basilar or anterior inferior cerebellar artery)

•Anterior tympanic branch of the maxillary artery

•Stylomastoid branch of the posterior auricular artery

•Petrosal branch of the middle meningeal artery



[Peterson, 2020]

Management:

Acute management: [maintain airway and oxygenation]

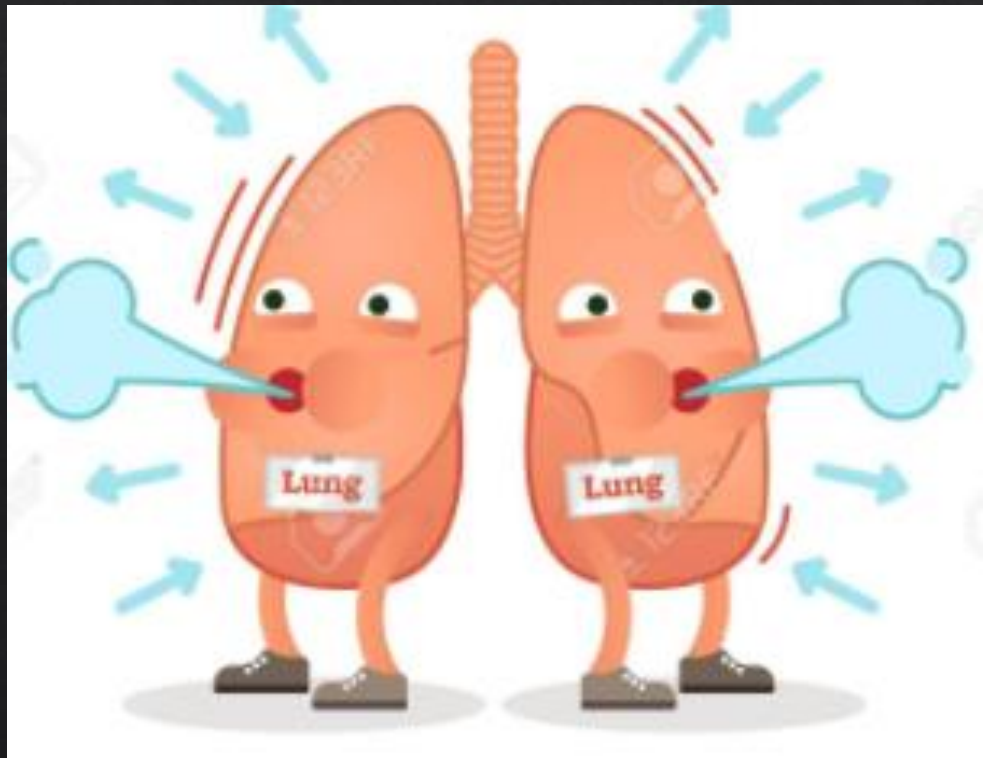
- Mechanical ventilation:
  - Apneustic and pneumotaxic center in pons impaired
  - Vitals (BP, HR)

Treat the underlying cause immediately:

- Ischemic stroke
  - Thrombolytic therapy

Rehabilitation

- Speech therapist
- Occupational therapist
- Physical therapy



Pulmonary complications represented the leading cause of death at first week post onset of the "locked-in" state.

(Patterson, 1985)

- In a study it was found that mean time to diagnosis of LIS was 78.8 days.
  - Transitioning from coma→ LIS

- More than 50% of the time, the caregiver notices subtle movements which prompts workup
  - When suspicious for LIS→ MRI & MRA for dx

Prognosis:

If the cause is non-vascular→ moderate to full recovery

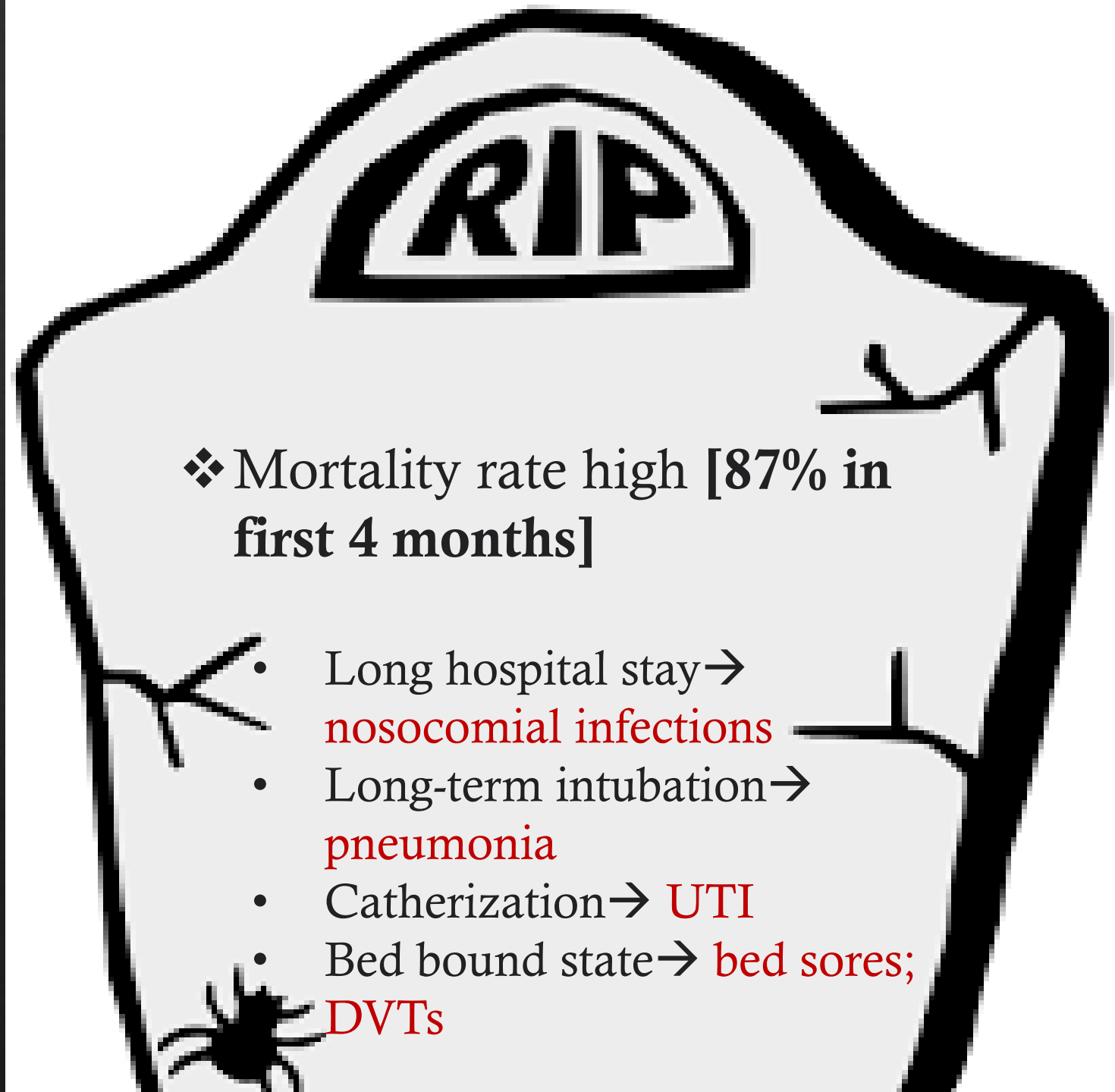
If the cause is vascular→

- No recovery [no return of motor function]
- Minimum recovery[some voluntary motor return but fully dependent on other for care]
- Moderate recovery [noticeable return of motor function; complete some but not all activities during daily living
- Full recovery→ regained ability to do all activities; slight neurological deficits
- No neurological deficit

Life with LIS:

- One study reports 18/29 cases survived; 11-residing in own home requiring some assistance; others living in permanent hospital residents and nursing homes.

- Few go on to live independent lives with a job



- Mortality rate high **[87% in first 4 months]**

  - Long hospital stay→ nosocomial infections
  - Long-term intubation→ pneumonia
  - Catherization→ UTI
  - Bed bound state→ bed sores; DVTs

Long term survival in LIS patients?

10 years→ 83%

20 years→ 40%

Quality of life:

- In 2007, questionnaire was sent to 197 people , of which 67 patients responded with a completed questionnaire, of which 39 returned questionnaire in 2013.

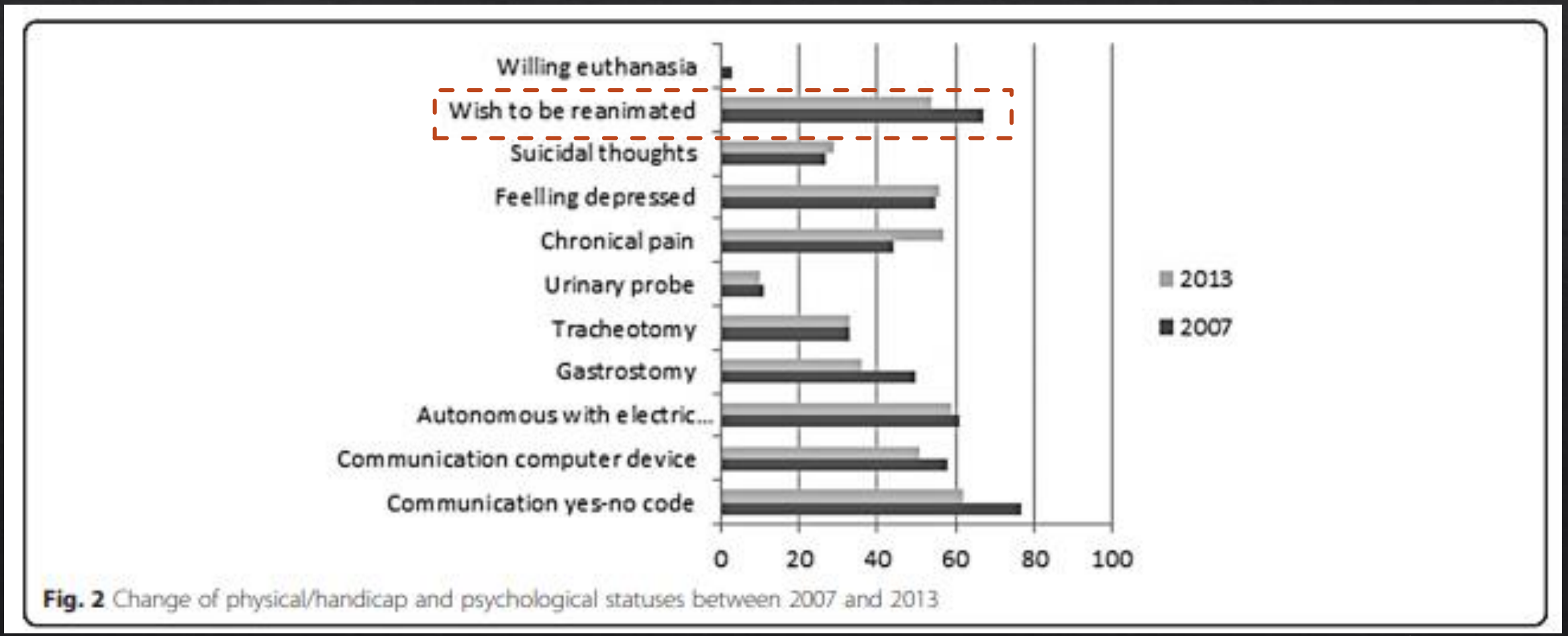


Fig. 2 Change of physical/handicap and psychological statuses between 2007 and 2013

(Rousseau, 2015)

Differentials:

Persistent vegetative state:

- Not conscious
-No voluntary motor movements
- respiration intact
-Preserved eye opening

Akinetic mutism:

-Preserved consciousness
-Brainstem reflexes intact
-Slow movements (no paralysis)
-Slow speech

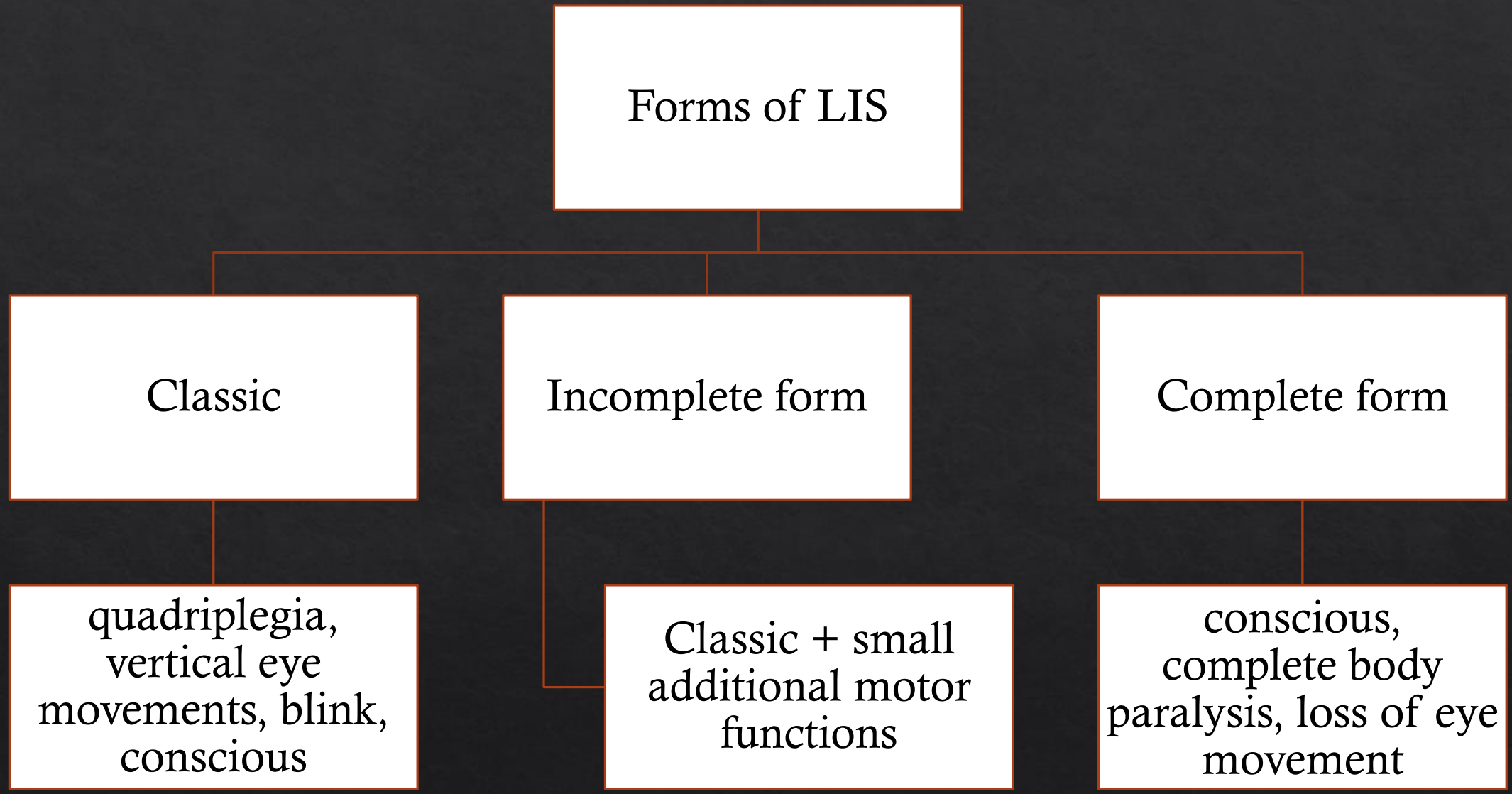
Coma:

- Not conscious
- No voluntary motor movements
-brainstem reflex varies
-respiration varies

Brain death:

-Not conscious
-No voluntary motor movements
-No brainstem reflexes
-No respiration

(Das, 2020)


Forms of LIS
Classic
Incomplete form
Complete form
quadriplegia, vertical eye movements, blink, conscious
Classic + small additional motor functions
conscious, complete body paralysis, loss of eye movement

(Das, 2020)

“It felt like I was in a really bad nightmare constantly for about the first three months. I could only just hear (I couldn't even open my eyes or breathe by myself); without them even knowing that I still could hear, the doctors and specialists in front of me said to my mum that I would die. They even asked my mum if she wanted them to turn the life support machine off after a few days.”

- Patient x

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That's all folks!