

A 10-year-old boy is brought to the clinic with headache, vomiting, and visual disturbances. The patient has had 2 weeks of persistent headache for which he was given over-the-counter analgesics without relief. He has also had daily episodes of nonbloody, nonbilious emesis. Neurologic examination shows bilateral upper eyelid retraction and limitation of upward gaze with a preference for downward gaze. Pupils react sluggishly to light but respond appropriately to accommodation. Strength, sensation, and deep tendon reflexes are normal. Plantar reflexes are downgoing bilaterally. Which of the following lesions is most likely responsible for this patient's symptoms?

- ☐ A. Craniopharyngioma
- ☐ B. Medulloblastoma
- ☐ C. Neuroblastoma
- ☐ D. Pinealoma
- ☐ E. Retinoblastoma

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- ☐ A. Craniopharyngioma [23%]
- ☐ B. Medulloblastoma [17%]
- ☐ C. Neuroblastoma [4%]
- ☒ D. Pinealoma [54%]
- ☐ E. Retinoblastoma [2%]

[Proceed to Next Item](#)**Explanation:**User Id: XXXXXXXXXX

Clinical features of pineal gland masses	
Parinaud syndrome (dorsal midbrain syndrome)	<ul style="list-style-type: none">• Limited upward gaze• Upper eyelid retraction (Collier sign)• Pupils non-reactive to light, reactive to accommodation
Obstructive hydrocephalus	<ul style="list-style-type: none">• Papilledema• Headache, vomiting• Ataxia

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This patient's history and physical examination are concerning for a **pineal gland tumor**, or pinealoma. The **pineal gland** is located in the quadrigeminal cistern and is

hydrocephalus

- Headache, vomiting
- Ataxia

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This patient's history and physical examination are concerning for a **pineal gland tumor**, or pinealoma. The **pineal gland** is located in the quadrigeminal cistern and is responsible for melatonin production. Pineal gland tumors are rare, and germ cell tumors account for the majority of occurrences. Any abnormal pineal growth can produce serious complications from mass effect (eg, **Parinaud syndrome**, **obstructive hydrocephalus**).

Parinaud syndrome results from pressure on the pretectal region of the midbrain near the superior colliculus and cranial nerve III. Classic examination findings include **limitation of upward gaze** with a downward gaze preference, bilateral **eyelid retraction** (eg, Collier sign, sclera visible above the superior corneal limbus), and **light-near dissociation** (eg, pupils that react to accommodation but not to light).

Pineal gland masses can also block cerebrospinal fluid flow in the aqueduct of Sylvius, causing obstructive hydrocephalus and symptoms of headache and vomiting.

(Choice A) Craniopharyngiomas are suprasellar masses that can compress the optic chiasm and result in visual field deficits but are not associated with Parinaud syndrome.

(Choice B) Pediatric **medulloblastomas** (red circle) usually arise from the cerebellar vermis and present with ataxia and truncal instability from hydrocephalus (red arrow) and mass effect. Medulloblastomas are not associated with Parinaud syndrome.

(Choice C) Neuroblastomas arise from the sympathetic nervous system and are usually located in the adrenal glands; intracranial tumors are rare. Opsoclonus myoclonus syndrome (antibody-mediated "dancing eyes and feet") can occur but is not associated with Parinaud syndrome.

(Choice E) Retinoblastoma is an intraocular tumor most common in children age <5. A "trilateral" retinoblastoma consists of bilateral retinoblastoma and a pineal gland tumor. It is rare, especially in an older child, and typically presents with **leukocoria**.

Educational objective:

Parinaud syndrome occurs in most patients with pineal gland masses and presents as limitation of upward gaze, bilateral eyelid retraction, and light-near dissociation. Pineal gland tumors can also present with headache and vomiting due to obstructive hydrocephalus.

References:

1. Teaching neuroimaging: dorsal midbrain (Parinaud) syndrome with

serious complications from mass effect (eg, **Parinaud syndrome**, **obstructive hydrocephalus**).

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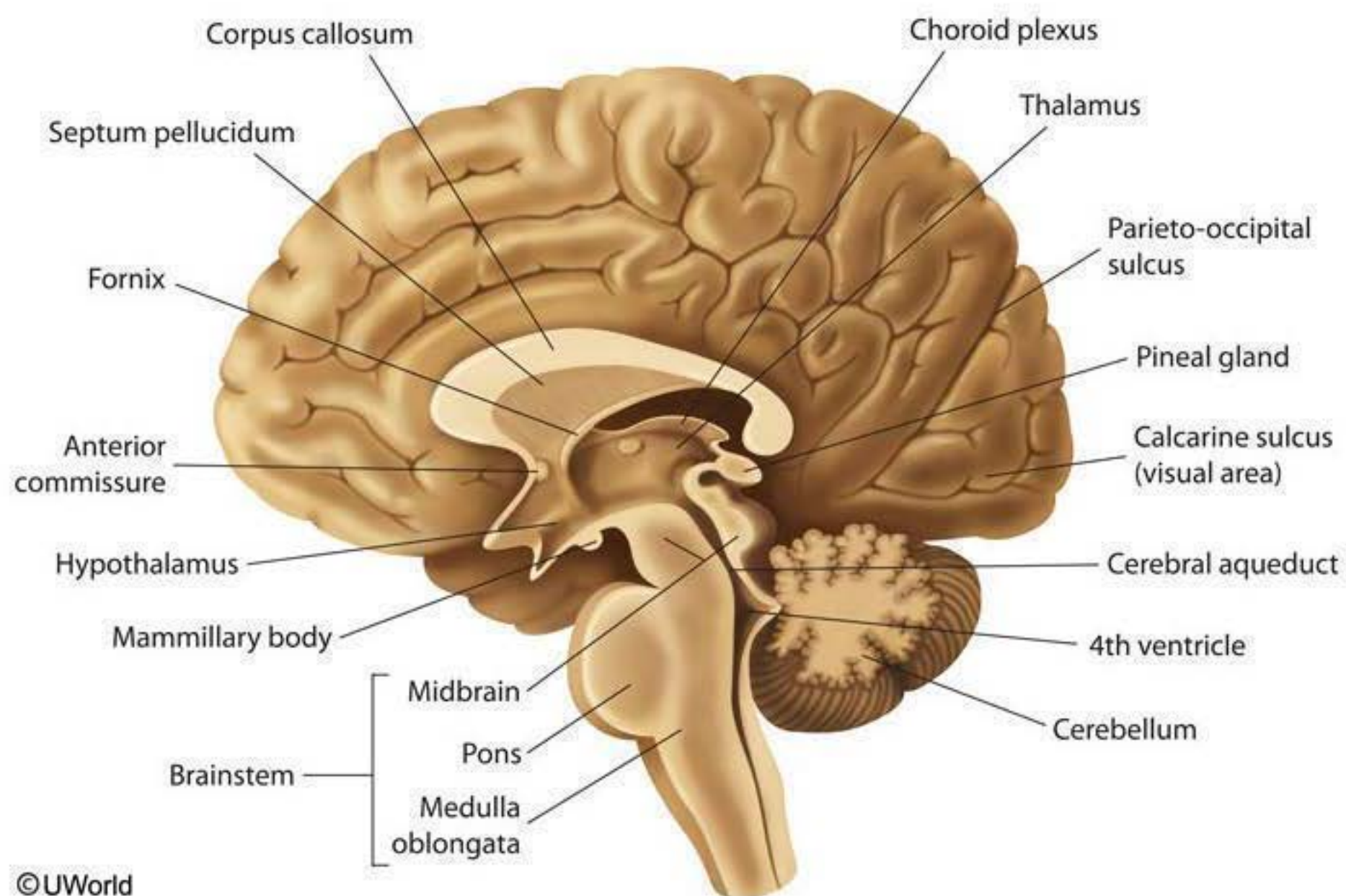
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References:

1. **Teaching neuroimages: dorsal midbrain (Parinaud) syndrome with corectopia.**
2. **Masses of the pineal region: clinical presentation and radiographic features.**
3. **Pineal region tumors in children.**

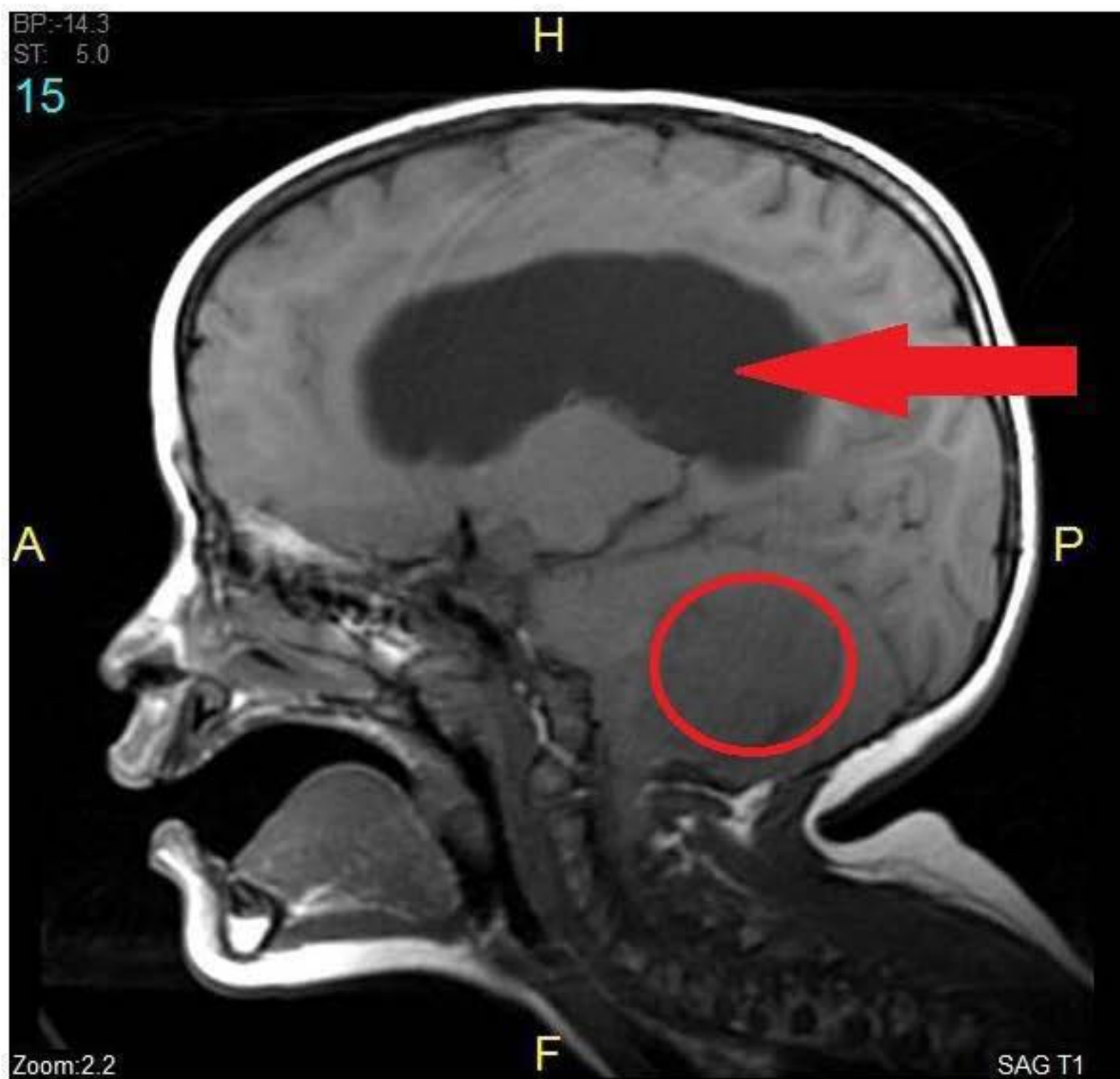
Media Exhibit

brain



Media Exhibit

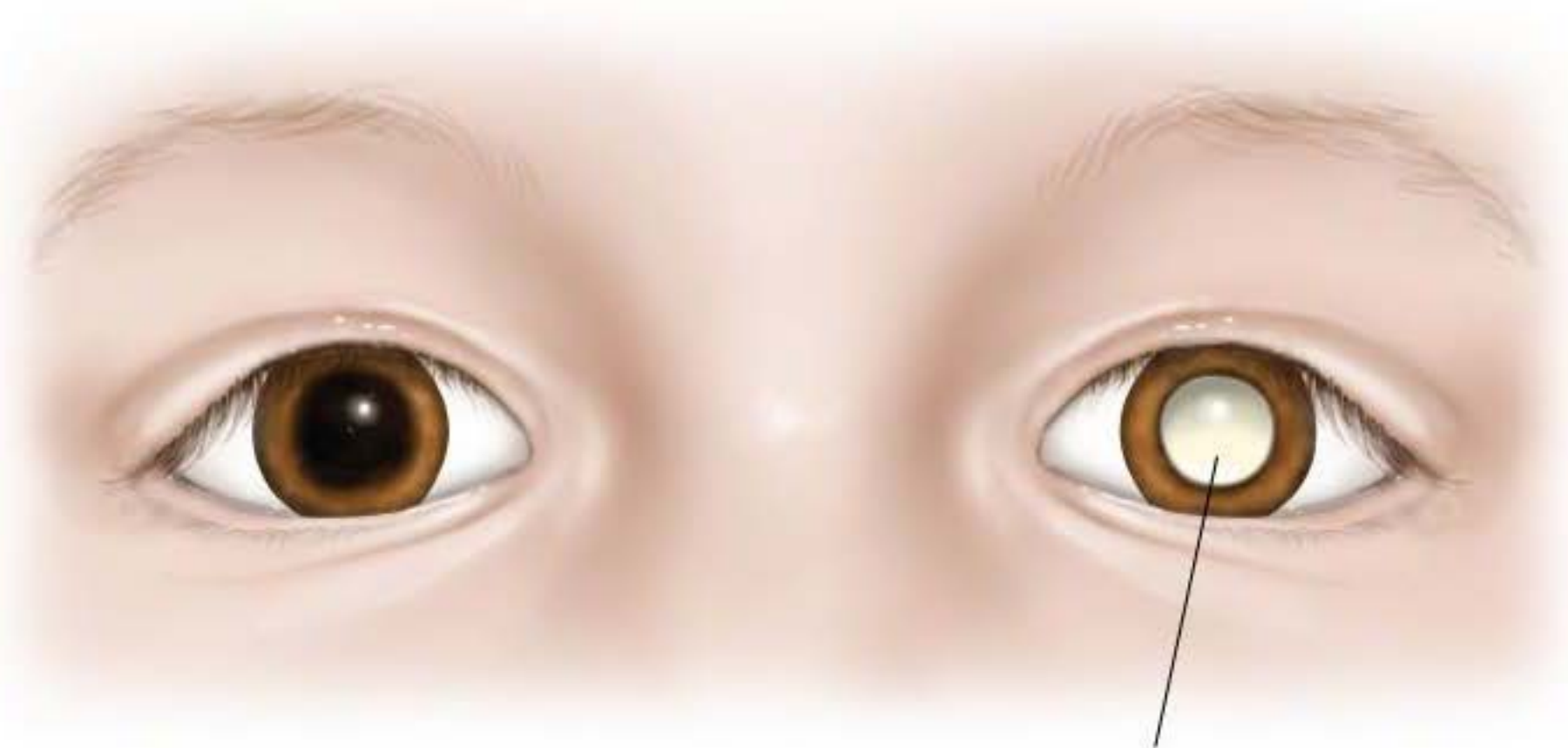
blastoma



Media Exhibit

Retinoblastoma white reflex

Retinoblastoma white reflex



Retinoblastoma of the left
eye with white reflex

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