

A 5-year-old boy is brought to the office by his parents for evaluation of "clumsiness." The patient's fine and gross motor development have been normal at prior visits, but he now frequently runs into doorways or corners of furniture while walking around the house. He has also begun drinking large amounts of water and urinating "all the time." Temperature is 37 C (98.6 F), blood pressure is 95/65 mm Hg, and pulse is 130/min. Physical examination shows dry mucous membranes and loss of bitemporal visual fields. Laboratory results are as follows:

Serum studies

Sodium	145 mEq/L
Potassium	3.9 mEq/L
Bicarbonate	20 mEq/L
Blood urea nitrogen	18 mg/dL
Creatinine	0.9 mg/dL
Blood glucose	88 mg/dL

Urinalysis

Specific gravity	1.001
Glucose	negative

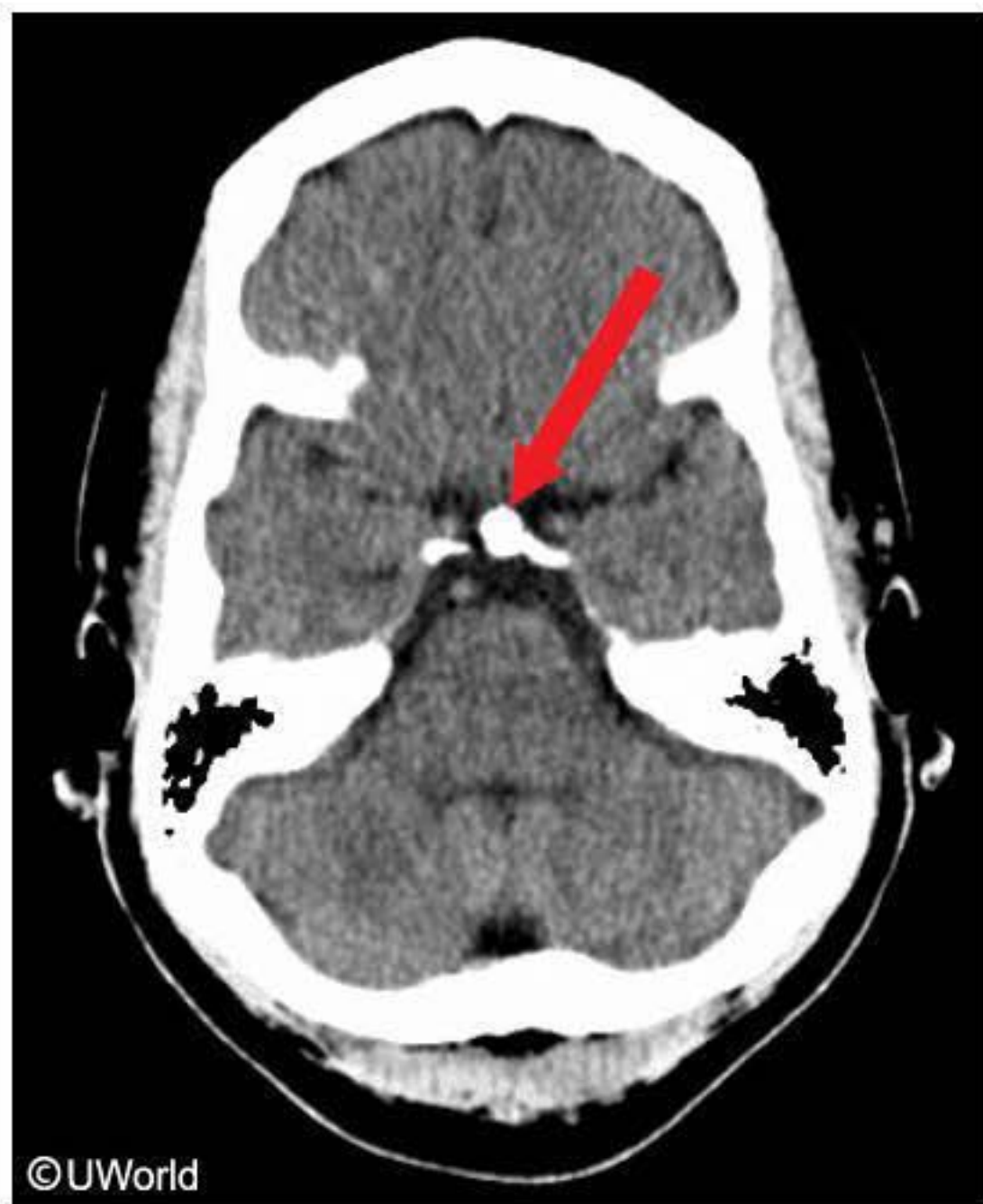
CT scan of the brain ([exhibit](#)) reveals an abnormality (red arrow). Which of the following is the most likely diagnosis?

- ☐ A. Craniopharyngioma
- ☐ B. Empty sella syndrome
- ☐ C. Medulloblastoma
- ☐ D. Pituitary adenoma
- ☐ E. Rathke cleft cyst
- ☐ F. Retinoblastoma

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- ☒ A. Craniopharyngioma [69%]
- ☐ B. Empty sella syndrome [2%]
- ☐ C. Medulloblastoma [1%]
- ☐ D. Pituitary adenoma [22%]
- ☐ E. Rathke cleft cyst [5%]
- ☐ F. Retinoblastoma [0%]

is the most likely diagnosis?

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Explanation:

User Id: [REDACTED]

Key features of craniopharyngioma

- Low-grade malignancy derived from remnants of Rathke pouch
- Optic chiasm compression → bitemporal hemianopsia
- Pituitary stalk compression → endocrinopathies (eg, growth hormone deficiency, diabetes insipidus)
- Suprasellar, calcified mass on imaging

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Craniopharyngiomas are low-grade malignancies derived from epithelial remnants of Rathke pouch within the pituitary stalk and occur in the **suprasellar** region adjacent to the optic chiasm. Compression of the optic chiasm can lead to visual field deficits including **bitemporal hemianopsia**, which may cause patients to run into the corners of walls or furniture. Pituitary stalk compression can lead to multiple **endocrinopathies**, including growth hormone deficiency and diabetes insipidus (eg, polyuria/polydipsia, high-normal serum sodium, dilute urine).

As with most intracranial tumors, craniopharyngiomas can also present with headache secondary to compression of nearby structures or obstructive hydrocephalus.

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As with most intracranial tumors, craniopharyngiomas can also present with headache secondary to compression of nearby structures or obstructive hydrocephalus. Craniopharyngiomas can be seen on MRI and CT scan, with **calcifications** in the vast majority of **lesions** (red arrow).

(Choice B) In **empty sella syndrome**, the sella turcica is often enlarged (red arrow) and contains no discernible pituitary gland (yellow arrow). Patients may be asymptomatic with an empty sella as an incidental finding on imaging or present with hypopituitarism but would not have calcifications.

(Choice C) Medulloblastomas are the second most common tumor of the posterior fossa following astrocytomas in children. These tumors typically arise from the cerebellar vermis and present with vomiting, headache, and ataxia. Calcifications are not typically seen.

(Choice D) Pituitary adenomas are benign neoplasms of the anterior pituitary with a portion of these presenting with manifestation of pituitary hormone secretion (eg, prolactinoma with galactorrhea and amenorrhea). Patients with pituitary adenomas can also present with bitemporal hemianopsia but would not have calcifications within the mass.

(Choice E) Rathke cleft cysts are also derived from Rathke pouch remnants. If large, their symptoms can be similar to those seen in craniopharyngiomas, but Rathke cleft cysts occur in the sellar region and are not typically associated with calcifications.

(Choice F) Retinoblastomas are the most common pediatric intraocular tumor and classically present with leukocoria or a white reflection in place of the standard **red reflex**.

Educational objective:

Craniopharyngiomas are calcified, intracranial tumors that occur in the suprasellar region. Presenting symptoms include bitemporal hemianopsia and pituitary hormonal deficiencies (eg, diabetes insipidus, growth hormone deficiency).

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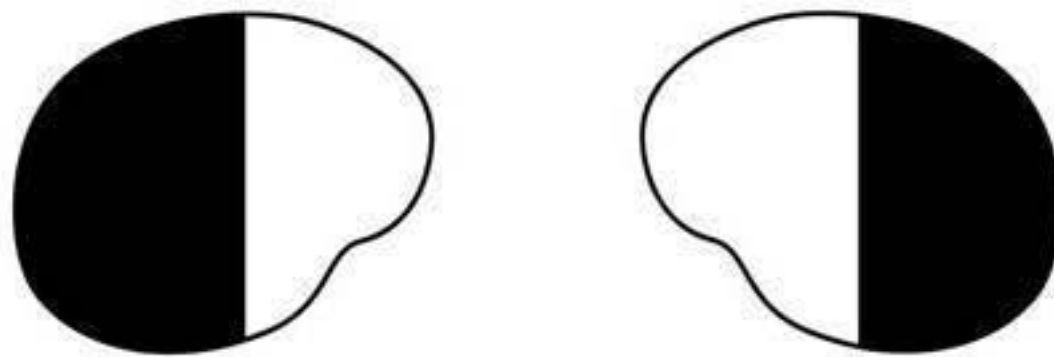
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References:

1. **Craniopharyngioma.**
2. **Craniopharyngioma.**

Media Exhibit

lateral hemianopsia



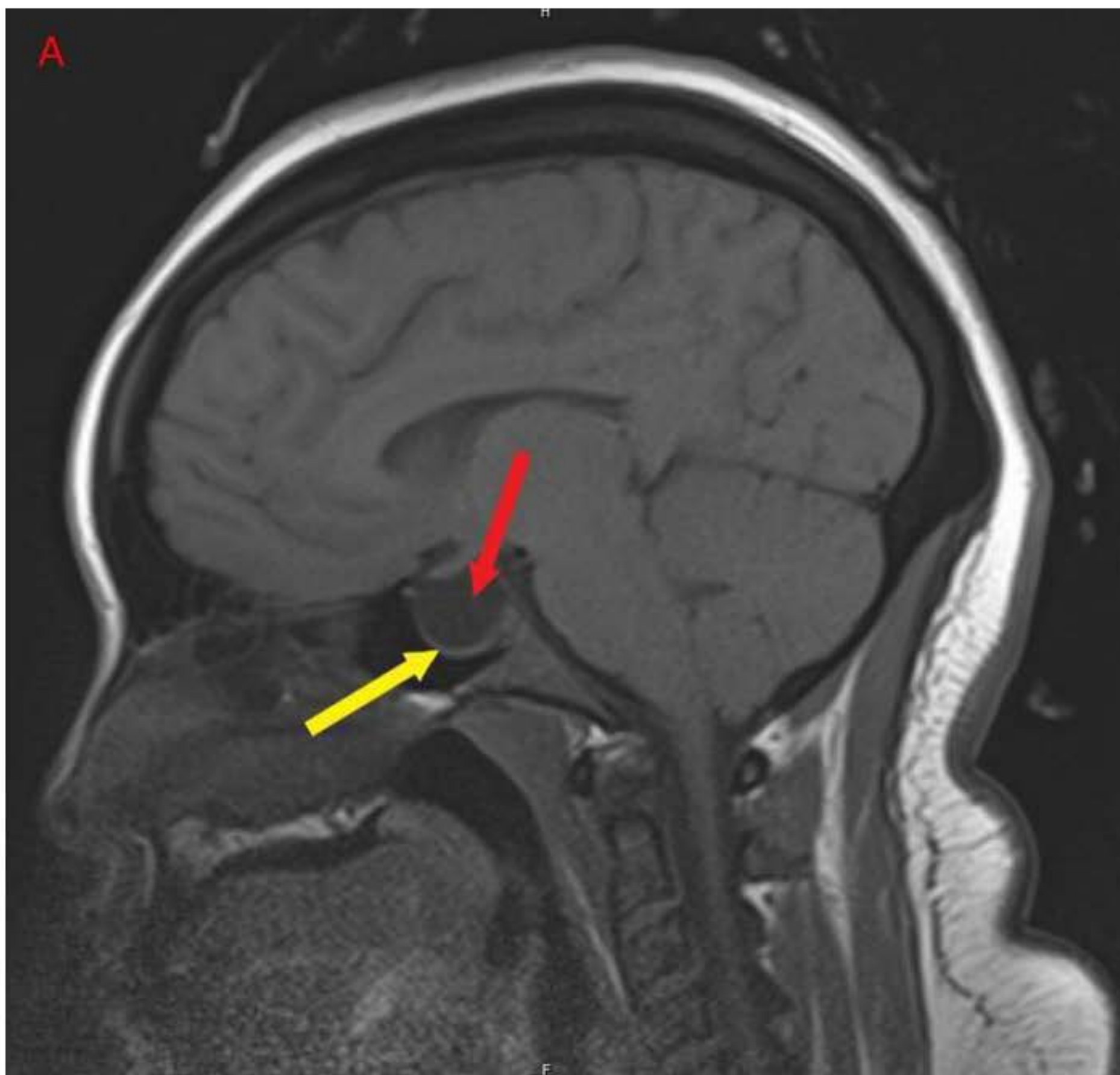
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Craniopharyngioma



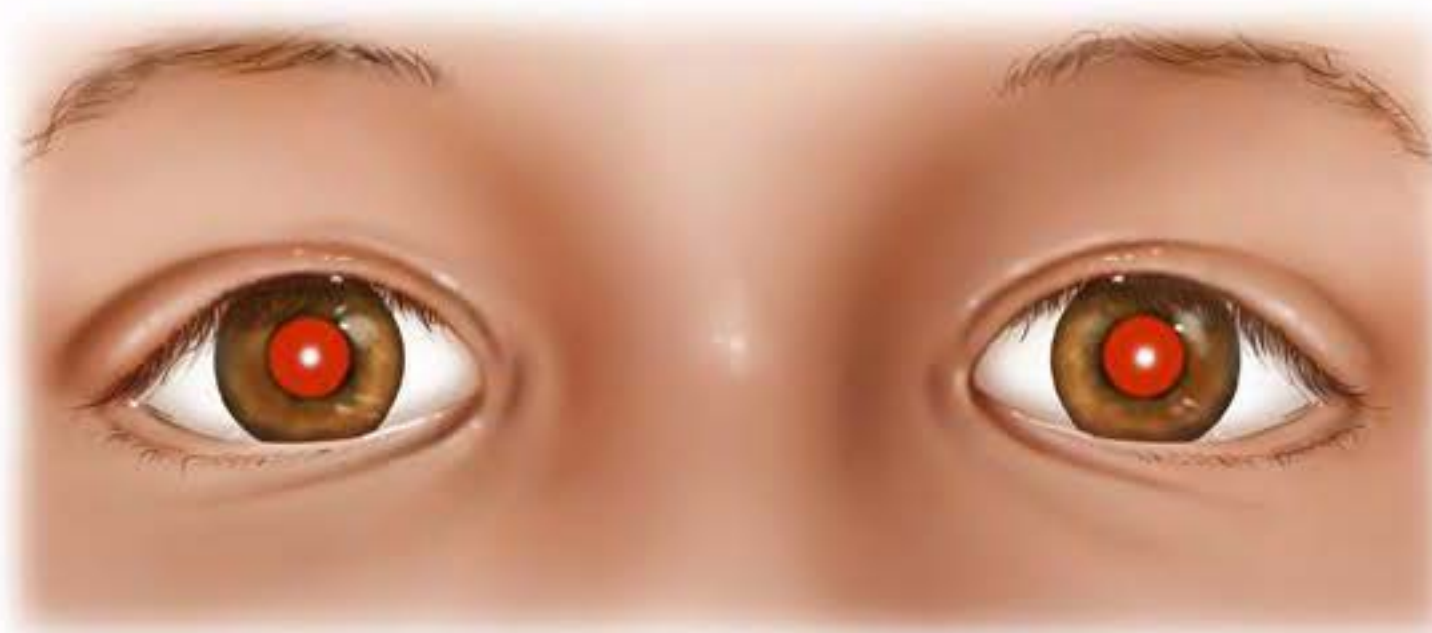
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ella syndrome



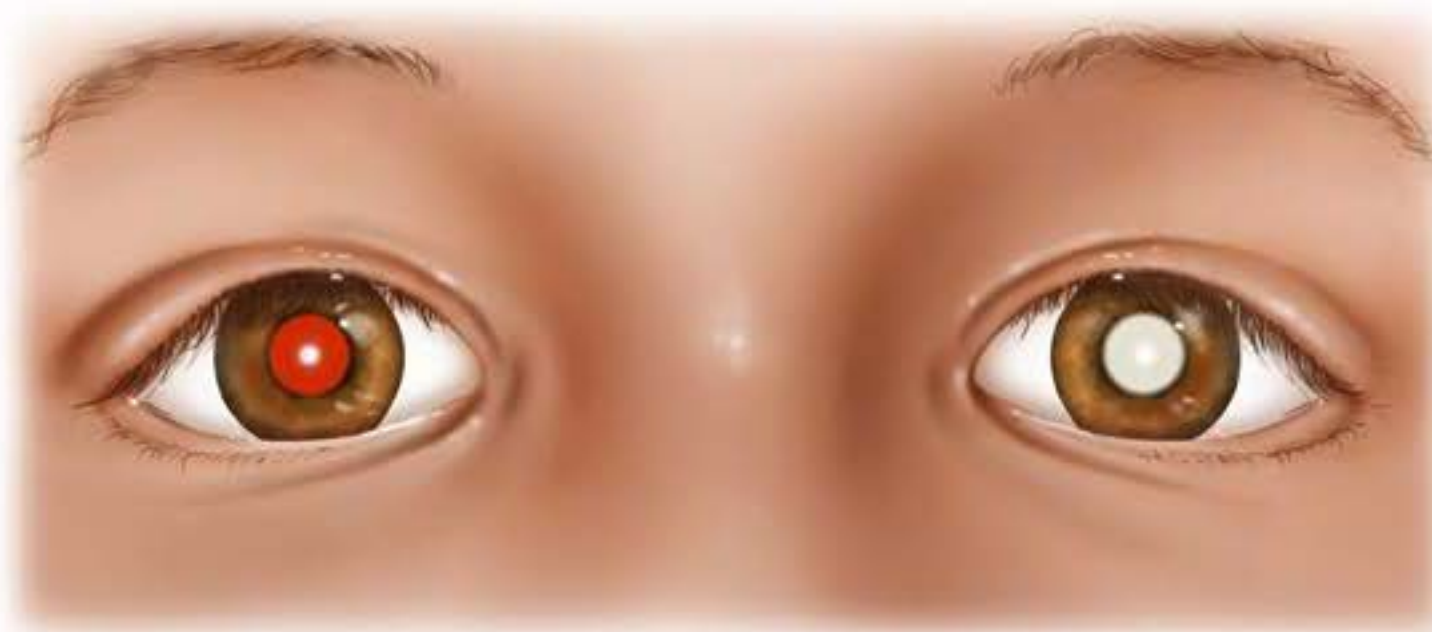
lex and corneal light reflection

Normal eyes & white reflex



Normal eyes

Red reflexes & corneal light reflexes are equal



Absent reflex

White reflex on abnormal eye can result from opacities of the lens (eg, cataract), or tumor (eg, retinoblastoma)

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