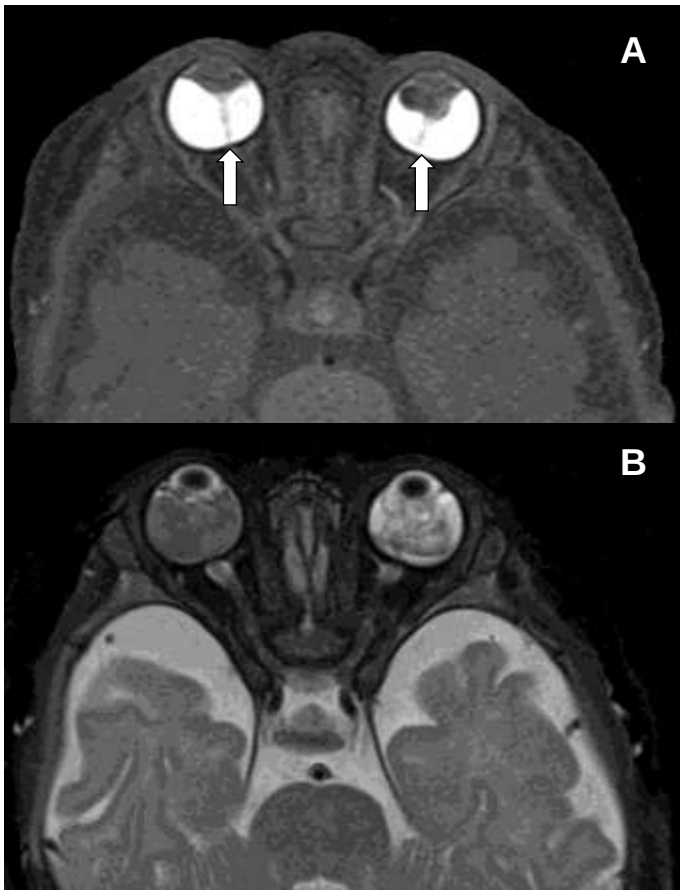
PERSISTENT HYPERPLASTIC PRIMARY VITREOUS (PHPV)



Case #6: A 6-month-old girl was brought in due to leukocoria in both eyes. Diagnosis?

PHPV is a spectrum of abnormalities caused by failure of involution of the embryonic hyaloid vascular system. It is usually unilateral but in this rare case, it is bilateral. After retinoblastoma, PHPV is the most common cause of leukocoria in childhood.

On MR, there is T1 hyperintensity of the vitreous chamber (*Figure A*). A low intensity signal linear structure extending from the posterior part of the lens to the optic nerve head (Cloquet canal, *arrows*) is seen. A retrolental mass can be present (*Figure B*), which usually enhances in the post-contrast study (*not shown*). Calcification is unusual.

On the other hand, retinoblastoma is a posterior globe mass usually containing calcifications. Coats disease (primary retinal telangiectasias) can mimic retinoblastoma, with a nonenhancing heterogeneous mass ± calcification. Retinal hemorrhage is likely also present in this case but as a complication.

References:

- Pediatric Radiology: Practical Imaging Evaluation of Infants and children. Edward Y. Lee, editor. Wolters Kluwer 2018.
- Caffey's Pediatric Diagnostic Imaging, 13th edition. Brian D. Coley, editor. Elsevier 2019.

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