

## Persistent Hyperplastic Primary Vitreous: Magnetic Resonance Imaging and Clinical Findings

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**Background:** This study presents magnetic resonance imaging (MRI) findings of both the anterior and posterior types of persistent hyperplastic primary vitreous (PHPV) to facilitate the differential diagnosis from other intraocular abnormalities.

**Methods:** Seventeen patients with PHPV who were evaluated using non-contrast and contrast-enhanced T1- and T2-weighted images were retrospectively reviewed.

**Results:** Among the 17 patients with PHPV (6 males, 11 females), 13 had unilateral PHPV (11 left eyes, 2 right eyes), and 4 had bilateral PHPV. The MRI findings of the anterior type of PHPV included a shallow or collapsed anterior chamber, an anterior segment anomaly, and a retrolental vascular membrane which demonstrated hyperintensity after contrast enhancement. The MRI findings of the posterior type consisted of microphthalmos; a tubular image, representing the hyaloid vessel; a funnel-shaped retinal detachment, with the subretinal fluid hyperintense on both T1- and T2-weighted images; the fluid-fluid level, which was hypointense on both T1- and T2-weighted images and probably corresponded to the presence of hemorrhage in the subretinal space; a retrolental mass; and vitreous hemorrhage. The most common clinical presentations of patients with PHPV in our study were microphthalmos, a shallow or collapsed anterior chamber, and leukocoria.

**Conclusions:** The presentation of PHPV at different stages was variable; the MRI features of PHPV along with clinical findings were able to facilitate the differential diagnosis from other intraocular abnormalities such as retinoblastoma and Coats' disease.

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**Key words:** persistent hyperplastic primary vitreous (PHPV), magnetic resonance image, retinoblastoma, Coats' disease.

Persistent hyperplastic primary vitreous (PHPV) is a unilateral, congenital malformation of the eye and is thought to be nonhereditary. This disorder is complicated by anterior and posterior chamber

anomalies and is clinically characterized by leukocoria, shallow anterior chamber, cataracts, angleclosure glaucoma, elongated ciliary processes, retrolental mass, microphthalmos, and retinal detachment. The

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primary vitreous forms around the seventh week of life and begins involuting at about 20 weeks.<sup>(1)</sup> PHPV arises as a result of the embryonic hyaloid vascular system failing to regress by the time of birth.<sup>(2)</sup> It may eventually lead to phthisis bulbi and intractable glaucoma. Moreover, intractable glaucoma and a clinically suspected retinoblastoma are the 2 main reasons for enucleation.<sup>(3)</sup> If the eyes have a cataract or opaque media and if direct visualization through an ophthalmoscope is impossible, more information to differentiate PHPV from a retinoblastoma can be obtained by using B-scan ultrasonography, computed tomography (CT), or magnetic resonance imaging (MRI); therefore unnecessary enucleation can be avoided. We report the MRI findings of PHPV and discuss their specific role in making a diagnosis of PHPV.

## METHODS

Seventeen patients with a diagnosis of PHPV were evaluated with MRI at Chang-Gung Memorial Hospital, Taoyuan from March 1992 to August 2000. In addition, systemic anomalies and other intraocular abnormalities associated with PHPV were also recorded. All patients underwent a complete ocular examination under general anesthesia in our outpatient department. The ocular examination included a slit lamp biomicroscopic examination with a KOWA portable slit lamp, tonometry examination with a tonopen, a direct or indirect ophthalmoscopic examination, color fundus photography, B-scan ultrasonography, and CT scan in some selected patients for detecting possible calcification when a retinoblastoma was suspected. After excluding a retinoblastoma, Coats' disease, and other intraocular tumors, data on 17 patients fulfilling the diagnosis of PHPV were collected. All orbital images in this study were evaluated in the axial and coronal planes with surface coils positioned to improve spatial resolution. The orbital T1-weighted images were obtained using the following parameters: repetition time (TR): 600-800 ms; echo time (TE): 20 ms; field of view, 12-16 cm; slice thickness, 3.0 mm; matrix, 256 × 192; and nose-to-ear-xiphoid index, 4. The orbital T2-weighted images were obtained using the following parameters: TR, 1800-2500 ms; TE, 20-80 ms; field of view, 16 cm; slice thickness, 3.0 mm; matrix, 256 × 192; and nose-to-ear-xiphoid index, 2. Signal inten-

sities of lesions were compared with that of the vitreous of the affected or opposite eye. Signal intensities were recorded as hyperintense, isointense, or hypointense on pre-contrast T1- and T2-weighted images. The degree of enhancement was evaluated on Gd-DTPA-enhanced T1-weighted images.

## RESULTS

There was a total of 17 patients including 6 males and 11 females. Unilateral PHPV was found in 13 (76.5%) cases, and among these, 11 (84.6%) involved the left eye and 2 (15.4%) involved the right eye (Table 1). Fourteen (82.4%) patients presented with microphthalmos, the most common sign in our study; a shallow or collapsed anterior chamber (76.5%), and with leukocoria (47.1%) as the second and third most common signs, respectively. In addition, glaucoma, cataracts, and strabismus were also relatively frequent. One bilateral case had retinal

**Table 1.** Demographics of Patients

Parameter	
Total no. of patients	17
Gender	
Male	6
Female	11
Unilateral	13
Left eye	11
Right eye	2
Bilateral	4
Age	5 w/o - 21 y/o

**Table 2.** Presenting Signs and Symptoms of Patients with PHPV

Signs and symptoms	No. of cases	Percentage
Microphthalmos	14	82.4%
Shallow AC	13	76.5%
Leukocoria	8	47.1%
Glaucoma	5	29.4%
Cataracts	5	29.4%
Strabismus	4	23.5%
Leukoma cornea	2	11.8%
Megalocornea	1	5.9%
Retinal hypoplasia	1	5.9%

**Abbreviations:** PHPV, persistent hyperplastic primary vitreous; AC, anterior chamber.

hypoplasia, and 1 case had buphthalmos (Table 2). We divided the MRI findings of PHPV into the anterior and posterior types (Table 3). Fourteen (82.4%) patients in our study had a combination of the anterior and posterior types, 1 (5.9%) patient had the pure anterior type, and 2 (11.8%) patients had the pure posterior type. The anterior type of PHPV included a shallow or collapsed anterior chamber, a retrolental vascular membrane, cataracts, and anterior segment anomalies. The retrolental vascular membrane was hyperintense after contrast enhancement (Fig. 1). A shallow anterior chamber, or microphthalmos, was easily detected when compared with the contralateral eye (Fig. 2). The posterior type of MR imaging find-

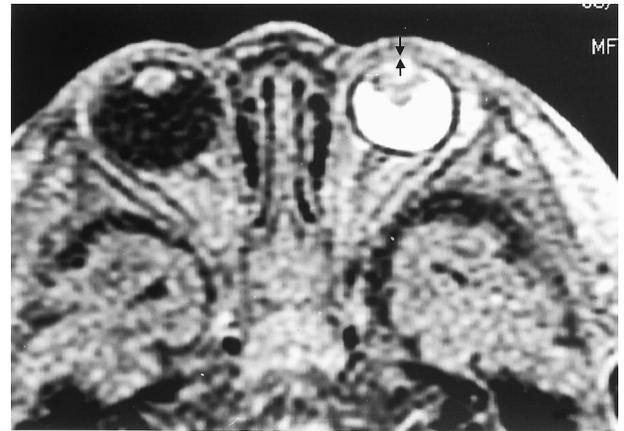
ings of PHPV included a tubular image (Fig. 3); a funnel-shaped retinal detachment, with the subretinal fluid appearing hyperintense on T1- and T2-weight-

**Table 3.** MRI Findings of PHPV

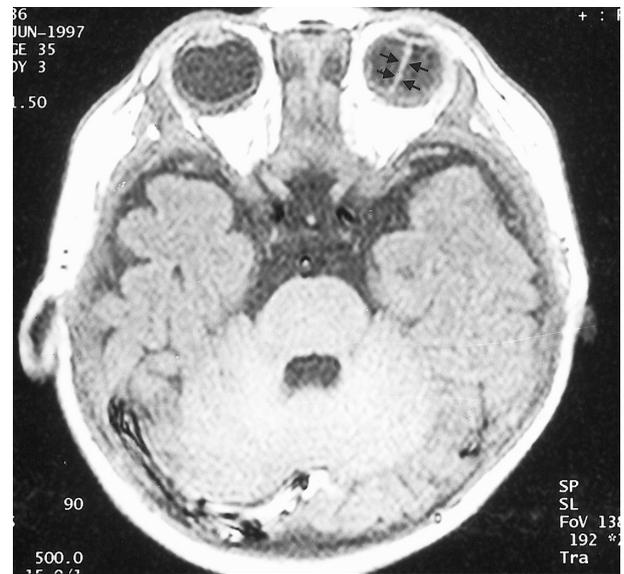
<b>Anterior type</b>	<b>( 5.9%)</b>
Shallow anterior chamber	
Retrolental vascular membrane	
Cataracts	
Anterior segment anomalies	
<b>Posterior type</b>	<b>(11.8%)</b>
Tubular image	
Funnel-shaped retinal detachment	
Fluid-fluid level	
Retrolental mass	
Vitreous hemorrhage	
<b>Combined type (anterior + posterior)</b>	<b>(82.4%)</b>



**Fig. 1** A 5-month-old girl with the anterior type of PHPV involving the left eye. An axial Gd-DTPA-enhanced T1-weighted MR image shows enhancement in the retrolental region (small arrow).



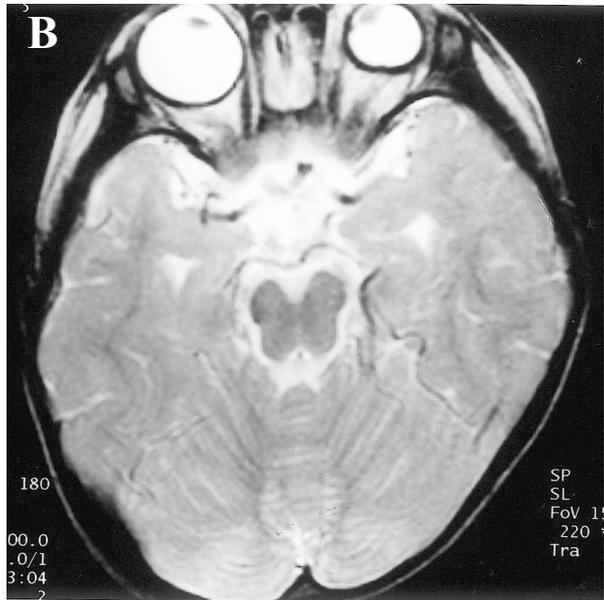
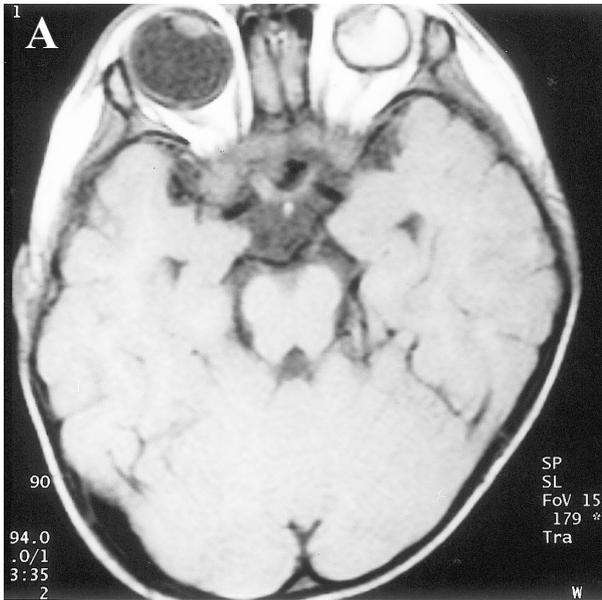
**Fig. 2** A 2-month-old girl with unilateral PHPV (os) involving the anterior and posterior chambers of the eye. An axial T1-weighted MR image shows a collapsed anterior chamber (small arrow) and microphthalmos compared with the normal right eye. In addition, funnel-shaped retinal detachment can also be noted.



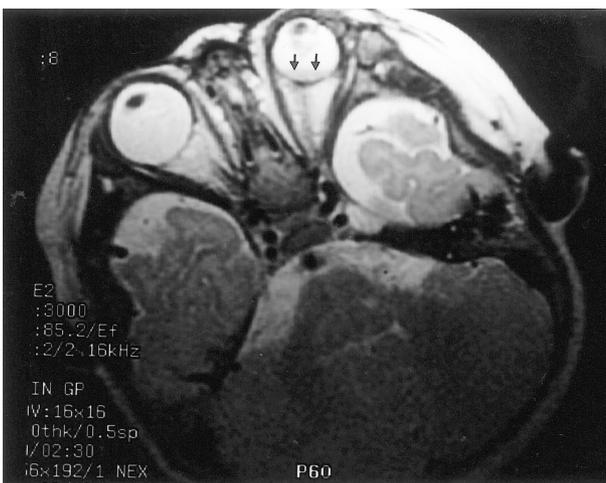
**Fig. 3** Axial T1-weighted image of both eyes. A 3-month-old boy had PHPV of the left eye. The involved eye has a normal-sized eyeball (compared with the contralateral eye). There is a tubular hypointense image in the left eye (small arrow).

ed images (Fig. 4); the fluid-fluid level, which demonstrated hypointensity on both T1- and T2-weighted images (Fig. 5); a retrolental mass (Fig. 6);

and vitreous hemorrhage, which was hypointense on the T1-weighted image (Fig. 7).



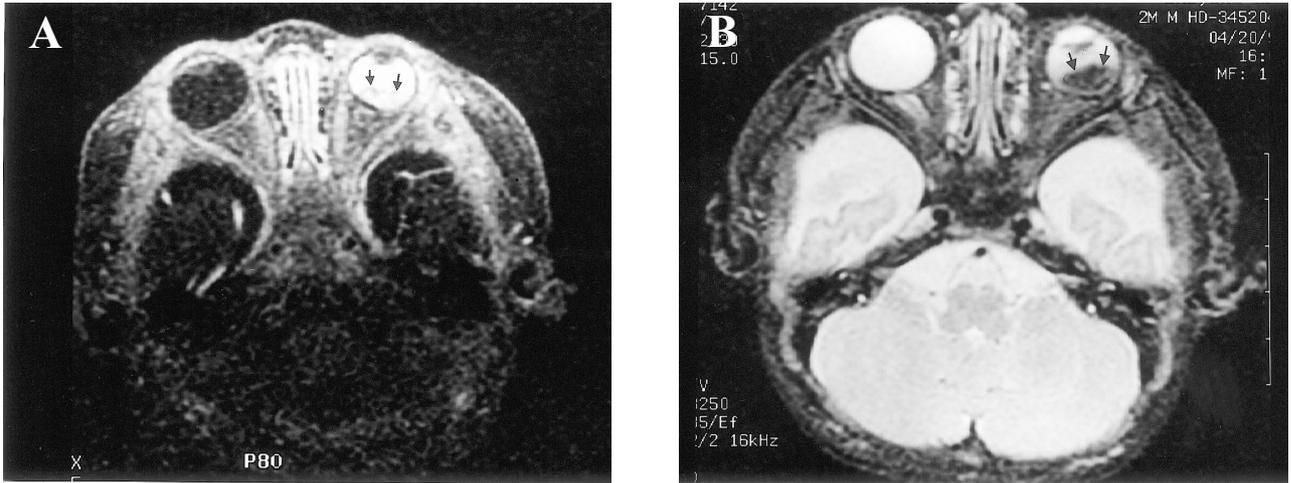
**Fig. 4** PHPV of the left eye. (A) An axial T1-weighted MR image. The left eye developed a funnel-shaped retinal detachment. The subretinal fluid was hyperintense. (B) An axial T2-weighted MR image of the same patient. Note that the subretinal fluid is also hyperintense.



**Fig. 5** Axial T2-weighted image. The fluid-fluid level (small arrow) in the left vitreous chamber shows a hypointense signal. It may correspond to serosanguineous fluid or acute subretinal hemorrhage in the subretinal space.



**Fig. 6** Sagittal T1-weighted image of the left eye. A hypointense signal of the retrolental mass can be noted, which is continuous with a tubular image that represents the detached retina.



**Fig. 7** (A) A 3-month-old girl with PHPV of the left eye. A mass-like lesion can be noted in the vitreous chamber. It appears as a hypointense signal on the T1-weighted image. (B) A 2-month-old boy with PHPV of the left eye. A similar lesion with a smoother dome-shaped surface appears hypointense on the T2-weighted image. These are considered to be vitreous hemorrhage.

## DISCUSSION

The fetal intraocular vascular system can be divided into anterior and posterior systems. The anterior system supplies the iris anterior to the lens, while the posterior system consists of 3 components: the main hyaloid artery which mainly supplies the central primary vitreous; the vasa hyaloidea propria, which supplies the peripheral portion of primary vitreous; and the tunica vasculosa lentis which supplies the iris and lens.<sup>(1,4)</sup> The primary vitreous lies between the lens and the retina. It contains the hyaloid vessels and fibrillar ectodermal tissues, then gradually disappears in the fifth to sixth month of gestation and is replaced by the secondary vitreous, which is the final vitreous. The anterior and posterior hyaloid vascular systems regress independently. If they fail to regress normally, then PHPV occurs. PHPV, therefore, may be clinically divided into a pure (anterior or posterior) type or a combined type.

Most cases of PHPV are unilateral, as in our study where 76.5% were unilateral and only 23.5% were bilateral. It is believed that bilateral cases are usually accompanied by systemic diseases such as Norrie's disease, Warburg's syndrome,<sup>(5)</sup> or other intraocular abnormalities.<sup>(3)</sup> One bilateral case in our study revealed retinal hypoplasia, intrauterine growth retardation (IUGR), and microcephaly, but the chro-

mosome study was normal. The common clinical presentations of PHPV included leukocoria, microphthalmos, an elongated ciliary process, retinal detachment, cataracts, glaucoma, and a shallow anterior chamber, but calcification was absent.<sup>(1,5)</sup> Microphthalmos, a shallow anterior chamber, and leukocoria were the most common presenting signs in this study.

Glaucoma is a relatively common complication, and it may be due to a closed angle, a pupillary block type, an immature filtration angle-induced or intumescence of the lens.<sup>(1,3)</sup> If left untreated, it will progress to intractable glaucoma, or buphthalmos, as presented in 1 of our cases. Previous studies revealed that PHPV is not hereditary, but 8 cases in 3 families were reported and showed bilateral persistence of the hyaloid artery.<sup>(6)</sup> In our study, there were 2 siblings from the same family, so PHPV is possibly heritable.

The CT and MRI findings of PHPV have been well studied,<sup>(1,4,5,7-10)</sup> but this is the first report to demonstrate the MR image findings of both the anterior and posterior types of PHPV. The first computed tomographic finding of PHPV was reported by Mafee<sup>(11)</sup> and Goldberg.<sup>(12)</sup> However, MR imaging is superior to CT in the diagnosis of PHPV, because it is valuable in revealing intraocular details and gravitational effects of the intravitreal fluid, and it is also

a better method to identify tissue components such as melanin, methemoglobin, deoxyhemoglobin, and proteinaceous fluid.<sup>(13,14)</sup> In addition, MR imaging subjects ocular tissues to no irradiation as compared with CT scans and has less biological side effects; it contributes to higher sensitivity and specificity in evaluating intraocular abnormalities.<sup>(2)</sup> Although B-scan ultrasonography and CT scans are easier to perform and are less expensive, the detection rate of calcifications on MR images is not as sensitive as on CT scans (MR images, 54%; CT, 82% as reported by De Potter et al.).<sup>(14)</sup> However, MRI still plays an important role in differentiating PHPV from noncalcified retinoblastomas and other intraocular abnormalities responsible for leukocoria, such as Coats' disease, Toxocara endophthalmitis, or retinal and vitreal disorders when CT scan and B-scan ultrasonography do not offer sufficient information. It was noted in this study that B-scan ultrasonography often made a limited contribution to a diagnosis of PHPV.

In our study, PHPV was divided into anterior and posterior types, with the majority of PHPV being the mixed type. The MRI findings of the anterior type included a shallow or collapsed anterior chamber, and a retrolental vascular membrane. This membrane appeared hyperintense after contrast enhancement. The finding of posterior type included the following. (1) Microphthalmos is an important finding to differentiate the posterior type from a retinoblastoma, because a microphthalmos is absent from eyes with a retinoblastoma. (2) A tubular image represents the hyaloid vessel or Cloquet's canal. (3) A funnel-shaped retinal detachment had subretinal fluid which appeared hyperintense on both T1- and T2-weighted images with respect to the vitreous of the contralateral eye. The high signal intensity was related to the presence of protein.<sup>(14)</sup> (4) The fluid-fluid level was revealed to be hypointense on both T1- and T2-weighted images compared with vitreous of the contralateral side and may correspond to subretinal hemorrhage. This appearance of hemorrhage depends on the level of oxygenation of the red blood cells. Deoxyhemoglobin is present first, followed by methemoglobin, and the breakdown of the blood ends up lysing the erythrocytes. At the deoxyhemoglobin stage, the hemorrhage is hypointense on T1- and T2-weighted images, but with development of erythrocyte lysis, the signal becomes increasingly hyperintense on T1- and T2-weighted images.<sup>(10,14-16)</sup>

(5) The retrolental mass also appeared tube hypointense on T1- and T2-weighted images. (6) Vitreous hemorrhaging was present. We found that the retrolental mass and funnel-shaped retinal detachment were the most common MRI findings of PHPV.

The differential diagnosis of PHPV includes retinoblastomas, Coats' disease, presumed ocular toxocariasis,<sup>(17-20)</sup> and some conditions contributing to subretinal fluid or hemorrhage. Coats' disease and retinopathy of prematurity (ROP) may have the same MRI findings as PHPV due to retinal detachment and subretinal fluid; however, retinoblastomas usually demonstrate hyperintensity on T1-weighted images and hypointensity on T2-weighted images. MRI findings.

Similar are choroidal melanomas, choroidal leiomyomas, choroidal metastasis, and medulloepitheliomas.<sup>(21)</sup> Factors contributing to retinal detachment are intraocular hemorrhage, incorporation of the peripheral retina into the retrolental mass, and retinal traction of the preretinal glial strand.<sup>(3)</sup>

In conclusion, unilateral involvement, microphthalmos, and a shallow or collapsed anterior chamber are the most important clinical clues in making the differential diagnosis of PHPV. When PHPV is suspected, a retrolental mass and a funnel-shaped retinal detachment are the most common MRI findings.

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## 持續存在之增生性初級玻璃體： 核磁共振掃描影像和臨床症候之表現

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- 背景：** 本文旨在提出持續存在之增生性初級玻璃體 (persistent hyperplastic primary vitreous) 其在眼睛前房及後房的核磁共振掃描影像之表現以便利於與其他易和此疾病混淆的眼內異常疾病相鑑別。
- 方法：** 蒐集17位有持續存在之增生性初級玻璃體並且做過核磁共振掃描的病人，紀錄其核磁共振掃描影像和臨床症候之表現。
- 結果：** 總共17個病人當中 (6位男性，11位女性)，有13位是單測發生 (11個在左眼，2個在右眼) 四位是雙側發生。持續存在之增生性初級玻璃體在眼睛前房的核磁共振掃描影像之表現包括有前房狹窄或塌陷、前房發育異常、及水晶體後之血管膜，此種血管膜加顯影劑後呈現高度顯影。而其在眼睛後房的影像表現有：一、小眼球；二、管狀影像，它代表玻璃狀體動脈；三、漏斗狀視網膜剝離，此視網膜下液體在T1及T2影像下呈現高度亮區；四、液狀介面，它在T1及T2影像下呈現暗區。它代表的是視網膜下出血；五、水晶體後腫塊；六、玻璃體出血。病人最常見的臨床表現是小眼球，前房狹窄或塌陷，以及白色瞳孔。
- 結論：** 持續存在之增生性初級玻璃體在不同階段其在眼睛內的表現呈現多樣化，而它在核磁共振掃描影像上的表現配合臨床上的表徵可以利於和其他眼內疾患如惡性視網膜母細胞瘤 (Retinoblastoma)、柯滋式病 (Coats's disease)等相鑑別。  
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**關鍵字：** 持續存在之增生性初級玻璃體，核磁共振掃描，惡性視網膜母細胞瘤，柯滋式病。

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