

# Ultrasonographic Feature of Persistent Hyperplastic Primary Vitreous

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## Abstract

**Purpose:** To observe the ultrasonographic features of patients with persistent hyperplastic primary vitreous (PHPV).

**Methods:** Thirty-two subjects (34 eyes) diagnosed with PHPV were evaluated by ultrasonography.

**Results:** The ultrasonography demonstrated a retrolental mass extending from the optic disc to the posterior lens capsule, manifested as band, regular triangle, or inverted triangle shapes. The band-shaped echo was characterized as a linear band extended from the optic disc to the posterior lens capsule. The regular triangle-shaped echo was manifested as a membranous septum with a wide base extended from the optic disc to the posterior lens capsule, and the anterior part became narrower. The inverted triangle echo was characterized as a membranous septum with a narrow base extended from the optic disc to the posterior lens capsule, and the anterior part become wider.

**Conclusion:** Ultrasonography is noninvasive and safe, and can offer real-time display of intraocular structures. This is especially important in individuals who are uncooperative or unsuitable for fundus examination due to media opacity. Combined with clinical feature, ultrasonography provides vital evidence for the diagnosis of PHPV. Thought observing ultrasonographic feature, clinicians could evaluate the size, position and severity of lesions in PHPV patients, and which would be helpful to determine the surgical approach and clinical

prognosis. (*Eye Science 2014; 29:100–103*)

**Keywords:** persistent hyperplastic primary vitreous; ultrasonography; feature

Persistent hyperplastic primary vitreous (PHPV), also known as persistent fetal vasculature (PFV)<sup>1</sup>, is a rare congenital malformation due to impaired regression of the hyaloid vascularization during the embryonic period. Clinical manifestations include leukocoria, microphthalmos, and anterior and/or posterior vitreous vascular residuals<sup>1,2</sup>. PHPV can be present in three forms: anterior, posterior and combined (both anterior and posterior). PHPV mainly occurs in children, lack of cooperation, and the clinical symptoms are atypical, so are likely to be misdiagnosed by fundus examination alone. Ultrasonography is safe and noninvasive, and real-time displays intraocular structures. In our study, 32 PHPV patients underwent ultrasonography and their ultrasonographic features are reported.

## Materials and methods

### Subjects

In total, 32 subjects (34 eyes) diagnosed with PHPV underwent ultrasonographies in our hospital and Sun Yat-sen University Cancer Center. between January 2008 and March 2014. The subjects included 21 males (23 eyes) and 11 females (11 eyes). The mean age was 7.23 years (range 3 months-44 years). Ten subjects (12 eyes) were aged  $\leq 1$  year old, 12 (12 eyes) aged 1-6 years, 6 (6 eyes) aged 6-18 years, and 4 (4 eyes) aged  $> 18$  years. Thirty PHPV cases (93.8%) were unilateral and 2 were bilateral. Clin-

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cal manifestations included failure to trace light and visual acuity decline in 13 cases, white spots in the eyes of 12 children found by their parents, strabismus in 4, photophobia in 2, and eyeball atrophy in 1. All subjects underwent comprehensive eye examinations including fundus evaluations after dilatation by ophthalmologists, and ultrasonographies. All of them were posterior or combined PHPV. Primary vitreous vascular residuals could be seen in front of optic disc and with/without lens opacity. All subjects were treated surgically and their clinical manifestations were further verified intraoperatively, confirming the diagnosis of PHPV<sup>1,3</sup>.

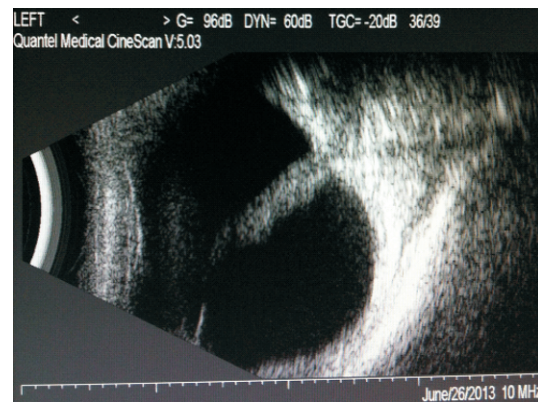
### Examination methods

The equipment utilized in this study was either an echographic examination with a 10 MHz probe (Quantel Medical CineScan, France) or a Philips iu22 ultrasound system device with L17-5 probe ultrasound transducer (iu22, Philips Medical Systems, Royal Philips Electronics, The Netherlands). Ultrasonography was performed in transverse and longitudinal planes while the child was lying in a supine position with both eyes closed. An abundant quality of gel was applied over the closed eyelid, and the hands of the examiner were placed on the borders of the orbits to avoid pressure on the globe and elevation of intraocular pressure. The uncooperative children took 10% chloral hydrate orally according to their body weight. Ultrasonography was performed as the route of lens-vitreous-retina-optic nerve in sequence. If any abnormal echo was detected, the position and morphology of echo and the relationship between the echo and the lens and optic nerve were carefully identified.

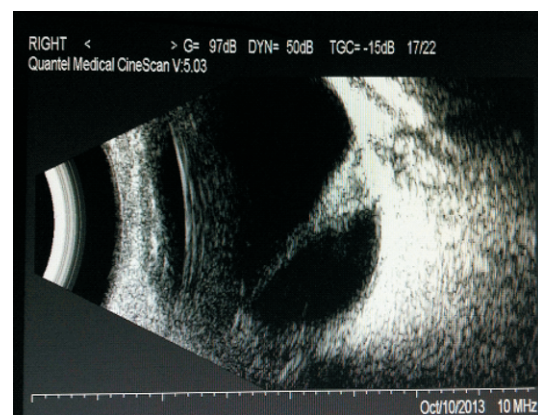
### Results

One key finding demonstrated by ultrasonography was a retrolental mass extending from the optic disc to the posterior lens capsule, and appeared as band, regular triangle, or inverted triangle shapes. Fifteen cases (17 eyes) had band-shaped echoes, presenting as the linear band extended from the optic disc to the posterior lens capsule, as shown in Figure 1. Regular triangle echoes were observed in 14 patients (14 eyes), manifested as the membranous septum with a wide base extended from the optic disc to the poste-

rior lens capsule, and the anterior part became narrower (Figure 2). Inverted triangle echoes were detected in 3 cases (3 eyes), manifested as the membranous septum with a narrow base extended from the optic disc to the posterior lens capsule, and the anterior part became wider, as illustrated in Figure 3. Movement of the retrolental mass was restricted to movement of the eye. Subjects with the complication of vitreous opacity showed intravitreal sporadic echo spots.



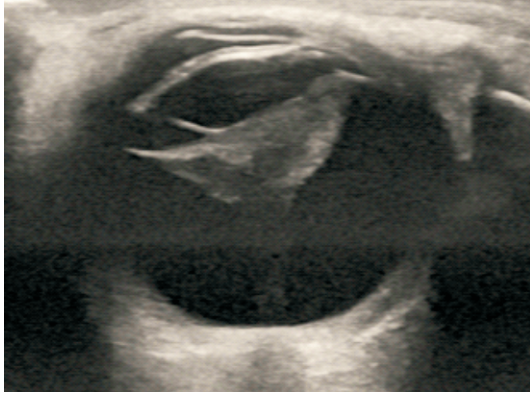
**Figure 1** The band-shaped echo was characterized as a linear band extended from the optic disc to the posterior lens capsule (left eye).



**Figure 2** The regular triangle-shaped echo was manifested as a membranous septum with a wide base extended from the optic disc to the posterior lens capsule, and the anterior part became narrower (right eye).

### Discussion

PHPV is a rare congenital malformation, resulting from impaired regression of the hyaloid vasculature during the embryonic period<sup>4</sup>. In this study, the per-



**Figure 3** The inverted triangle echo was characterized as a membranous septum with a narrow base extended from the optic disc to the posterior lens capsule, and the anterior part become wider(left eye).

centage of unilateral PHPV was 93.8% , consistent with previous findings<sup>5</sup>.

Routine examination of PHPV includes slit-lamp examination and ophthalmoscope examination. Ultrasonography is a noninvasive and highly reproducible method to evaluate the physical features, morphological structures, and functions of tissues, and it could also directly real-time display the position and morphology of lesions. It is valuable for research and is a useful auxiliary diagnosis tool in ocular diseases. Ultrasonography is convenient because measurements can be obtained through opaque media. It has vital application value in the diagnosis of PHPV, especially for uncooperative children or those with media opacity who are unsuitable for fundus examination.

Ultrasonographic outcomes in this study revealed three patterns: band, regular triangle, or inverted triangle shapes. Ultrasonography revealed that the band-shaped echo was characterized as a linear band extended from the optic disc to the posterior lens capsule. The inverted triangle echo was manifested as a membranous septum with a narrow base extended from the optic disc to the posterior lens capsule, and the anterior part become wider. The ultrasonographic features of these two echoes are consistent with previous findings<sup>6,7</sup>. We also observed a regular triangle-shaped echo, which manifested as a membranous septum with a wide base extended from the optic disc to the posterior lens capsule, and the anterior part became narrower. Sanghvi et al. also demonstrated a regular triangle echo but failed to provide

any further description or classification<sup>8</sup>.

We suppose that the three types of ultrasonography are correlated with the position and severity of lesions in PHPV patients. The posterior PHPV without anterior segment changes and the traction on the optic disc and retina is eliminated, or the combined PHPV with mild anterior segment changes and the traction on the optic disc and retina is weakened, thereby the ultrasonographies are manifested as band-shaped echoes. By contrast, the combined PHPV, which exerts strong traction on the optic disc and retina, and thereby alters their original positions, is characterized as a regular triangle echo in ultrasonography. The combined PHPV with severe anterior segment changes of a large sheet of retrolental fibrovascular membrane, which the traction on the optic disc and retina is not strong and the optic disc is not dislocated by external traction, ultrasonography is characterized as a large sheet echo posterior to the lens and a thin line shape echo anterior to the optic disc, like an inverted triangle shape. Previous studies merely described the ultrasonographic feature of PHPV but failed to further analyze the differences and the reasons. Ultrasonographic features and types of PHPV were contribute to the selection of surgical approaches and the prediction of postoperative effect. At present, we are conducting a prospective study to validate our hypothesis through preoperative ultrasonography.

Ultrasonography is safe and noninvasive, and reveals intraocular structures in a real-time manner. This procedure is especially useful for uncooperative patients or those with media opacity who are unable to undergo fundus examination. When combined with clinical characteristics, ultrasonography could offer pivotal evidence for the diagnosis of PHPV. Thought observing ultrasonographic feature, clinicians could evaluate the size, position and severity of lesions in PHPV patients, and which would be helpful to determine the best surgical approach and prediction of postoperative effect.

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