# Ultrasonographic evaluation of transition from normal to ectatic area: A comparison between myopic staphylomata and coloboma

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Choroidal coloboma and posterior staphyloma are two clinically distinct entities, with choroidal excavation being a unifying feature. They are associated with early onset cataract which can make ophthalmoscopy difficult. This report studies the transition between the normal and ectatic area in these cases with ultrasound. We evaluate "posterior hump" as a sign of differentiation between these two conditions.

**Key words:** Choroidal coloboma, ocular ultrasound, posterior staphyloma

Posterior staphyloma (PS) is *sine qua non* of pathological myopia.[1] It is characterized by disproportionate scleral ectasia with stretched conforming retinochoroidal layers. Conventionally, it has been classified into multiple types depending on location, although it usually involves the posterior pole of the eye. This outbulge, however, is a major risk factor for visually impairing maculopathy. PS can also be seen independent of high axial myopia.[2]

Choroidal coloboma (CC) is a developmental disorder, widely believed to be related to the defective closure of the embryonic optic fissure.[3-5] A typical CC is present in the inferior-nasal fundus, and severe forms (Types 1-3) also involve the posterior area.[6] It may not always be associated with its anterior counterpart: iris coloboma.[5] Like PS, it is characterized clinically by scleral ectasia. However, the overlying choroid is absent, and the retinal layers are defective and evidenced by an intercalary membrane.[7,8] Both hypermetropia and axial myopia may be associated, and unlike PS, CC may have systemic associations.[5]

Both PS and CC are associated with early onset nuclear sclerosis and cataract.[9,10] At such a stage, differentiating them on an ultrasound (USG)-based posterior segment evaluation (PSE) is challenging. We compare the USG findings of the transition zone from normal to ectatic area in cases with PS and CC and explore our findings as a new USG sign for differentiation.

## Methods

### Design

This is a masked prospective cross-sectional case-series. The study was conducted in accordance with the Declarations of Helsinki at a tertiary eye care center of northern part of India. Informed consent was obtained from all the participants. Consecutive patients with dense cataract sent for USG PSE to our dedicated ocular sonography services were included in the study. Fundus evaluation was not possible in these patients due to lenticular opacity. Patients who had undergone previous ocular/orbital surgery and those detected to have other ocular pathology on USG were excluded from the study.

For all the patients, USG was done by a single surgeon (AR), who was unaware of the clinical details. During the PSE, standard USG B-scan (EZ scan AB 5500+, Sonomed, NY., USA) was done and longitudinal and transverse scan images recorded. These eyes consequently underwent cataract extraction, and postoperative fundus evaluation for assessing the diagnoses was done by another surgeon (BT). A total of 27 eyes of 20 patients were detected to have diffuse choroidal

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excavations on USG due to CC, or myopic PS were included in the study. Later, all the USG images were independently evaluated for the presence of the USG sign by another surgeon (PV) who was masked to the clinical findings. Primary outcome measure was an analysis of the transition zone between the normal and ectatic area for the presence of a posterior hump [Fig. 1].

Data were tabulated on a Microsoft excel sheet and analyzed using SPSS version 22 software (SPSS Inc, Chicago, Illinois, USA). Clinical examination was considered as the gold standard and comparison was done for the presence of the USG sign between PS and CC with univariate Fischer's exact test. Sensitivity, specificity, predictive values and likelihood ratios were also calculated.

## Results

The mean age of the 20 patients was 44.89 + 12.70 years and 13 were male. A total of 27 eyes were included in the study, of which CC was noted in 12 eyes on postoperative fundus examination. Of these, iris coloboma was absent in five eyes (41.66%). A summary of the comparison between the two groups is presented in Table 1. The posterior hump was found to be present at the edge of all the cases of CC and in 13% of the cases of PS ($P = 0.0001$). Correct diagnoses of the choroidal excavation on the basis of USG had been made in 100% of the cases of CC and 87% of the cases of PS ($P = 0.49$).

The evaluation of posterior hump on USG as a diagnostic sign for CC is presented in Table 2. The sensitivity of the sign for diagnosing CC was found to be 100%, whereas it was found to be specific in 87% of the cases. The positive predictive value of the sign was 86%, whereas negative predictive value was 100%. The positive likelihood ratio was 7.5, whereas negative likelihood ratio was found to be 0. All these tests indicate the detection of posterior hump on USG to be a robust sign of CC.

**Table 1: Summary of comparison between the two Groups**

| | CC | PS |
|---|---|---|
| *n* | 12 | 15 |
| Mean age (years) | 39.75±16.10 | 49.0±7.43 |
| Male:female | 1:1 | 7:8 |
| Hump detected on USG ($P$=0.0001) | 12 | 2 |
| Correct diagnoses ($P$=0.49) (%) | 100 | 86.7 |

USG: Ultrasound, CC: Choroidal coloboma, PS: Posterior staphyloma

**Table 2: Evaluation of posterior hump as a diagnostic sign for choroidal coloboma on ultrasound**

| Diagnostic test | |
|---|---|
| Sensitivity (%) | 100 |
| Specificity (%) | 86.67 |
| Positive predictive value (%) | 85.71 |
| Negative predictive value (%) | 100 |
| Positive likelihood ratio | 7.50 |
| Negative likelihood ratio | 0 |

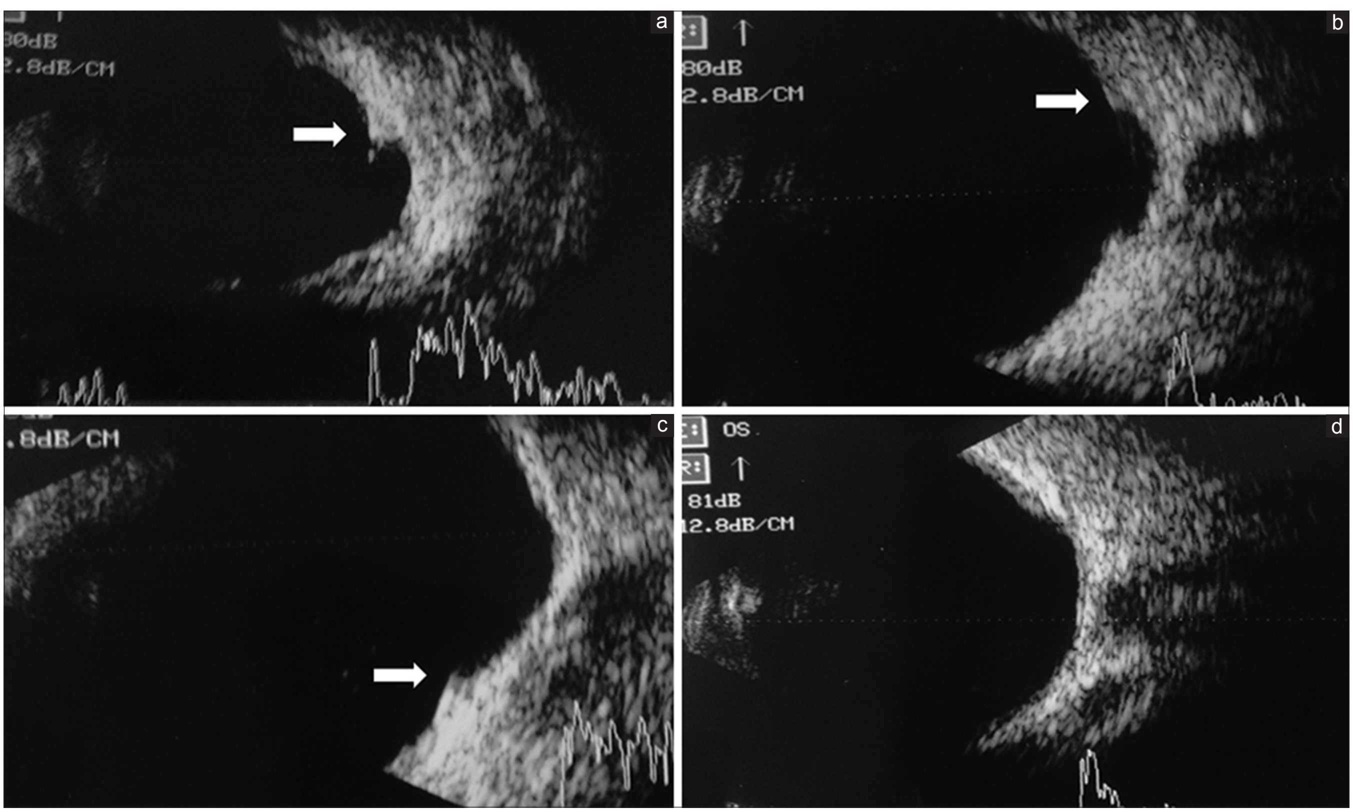


**Figure 1:** Ultrasound images of the posterior hump. (a-c) The posterior hump has been marked with white arrow at the edge of the choroidal excavation. (d) Large choroidal excavation can be seen without a posterior hump

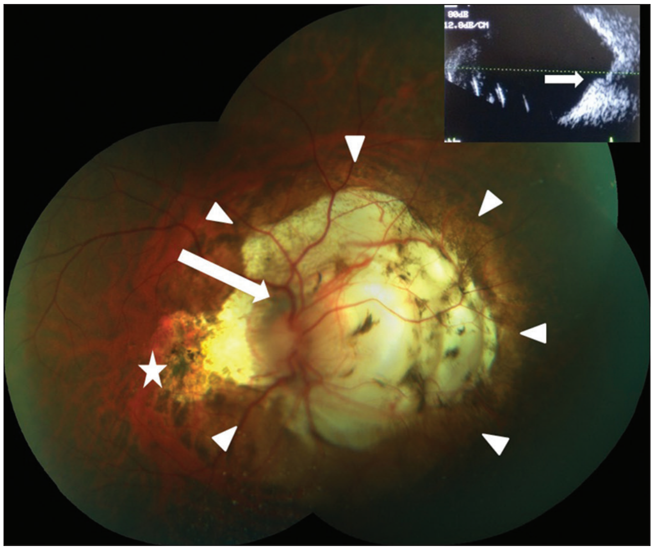

**Figure 2:** Fundus photo montage of an eye with posterior staphyloma detected to have the posterior hump. The border of the staphyloma can be visualized (white arrowheads) along with myopic maculopathy (white star). The optic disc is seen to be tilted (white arrow). Inset is the ultrasound image of this eye and the "pseudo-hump" has been indicated with white arrow

## Discussion

Gopal *et al*. have previously documented "the hump effect" on optical coherence tomography (OCT) at the edge of the CC.[7] They mentioned that "the eye wall was seen to make a sharp bend toward the vitreous cavity before merging with the coloboma." The author also stated that the hump was exaggerated due to the thickening of the retina. However, they detected this hump in only five of the 30 eyes, which is possibly due to the inclusion of eyes operated for retinal detachment and utilization of older OCT models (time-domain). In our patients, OCT imaging was not possible due to significant cataract. While a large number of retinal and choroidal changes have been described in eyes with PS using OCT, such humps have not been detected with either OCT B-scan or C-scan.[1,11] However, as OCT findings are typical of a subtle nature, comparing them with our USG findings may not be correct. The results of this study indicate an abrupt transition at the edge of coloboma-related ectasia as compared to a smoother transition at the edge of a myopic staphyloma. It should be noted that CC and iris coloboma can exist independently of each other and this USG sign is most relevant when clinical examination is not sufficient for differentiation.[5]

Two eyes with PS were also detected to have the posterior hump on USG. Both eyes were very long, more than 30 mm in axial length, and had severe chorioretinal atrophy in the area of the PS with an involved tilted optic disc [Fig. 2]. It seems that a combination of these findings led to detection of the "pseudo-hump." Nevertheless, severely long eyeballs with such distortion of the posterior pole should be kept as a less likely differential of this USG sign.

### Limitations

We included only two types of choroidal excavations and therefore, validation of this USG sign should be done by future larger-sized studies in a general setting. We chose myopic PS for comparison with CC as we needed a disease which did not have other apparent USG signs. As there are no other USG signs to differentiate CC from PS, Bayern's theorem cannot be applied to calculate posttest probability for this USG sign. Further, the impact of this difference between CC and PS on intraocular lens power calculation is beyond the scope of this study and needs evaluation.

## Conclusion

Detection of the posterior hump at the transition of normal to ectatic region on USG is useful for differentiating CC from PS in cases with insufficient optical media. Its utility in evaluating diffuse choroidal excavation should be evaluated with future studies.

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### Conflicts of interest

There are no conflicts of interest.

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