macular hole,[6] our study results suggest that residual hyaloid may not be a necessary factor for the development of ERMs in eyes with RD.

Finally, we concur with Robbie and Snead's opinion that the presence of Weiss ring is insufficient for the diagnosis of PVD. Importantly, the actual vitreoretinal relationship in the periphery is not known after the development of Weiss ring. Still, the appearance of Weiss ring remains the most easily identifiable and reliable sign to indicate that all or part of the vitreous hyaloid has been separated from the posterior retina.[3,4,6] Because we only used clinical examination for recruiting our cases, we specifically verified the presence of PVD by direct observation of the posterior vitreous during the initial phase of the surgery (see Materials and methods section in our article). In our conclusion, we used the phrase 'apparent PVD' to denote the inexactness of preoperative clinical examination in identifying true PVD.

We appreciate efforts put by the authors in the studies of the ultrastructures of vitreoretinal interface.[3,4] They raised the important issue of PHM, and its role in the formation of certain ERM. We hope further studies may clarify the significance of PHM in the formation of ERM after RD, and the interaction between TA and PHM.

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Eye (2007) **21,** 286–287. doi:10.1038/sj.eye.6702535; published online 4 August 2006

Sir,

Keratoconus in tuberous sclerosis

A 25-year-old Sri Lankan man presented with gradual deteriorating vision uncorrectable with spectacles. His best spectacle corrected visual acuity was 6/18 (right) and 6/9 (left), with no improvement with pinhole. Anterior segment examination showed Vogt's striae in both corneas. Fundoscopy revealed bilateral astrocytic hamartomas on the disc and nasal retinal hamartoma in the right (Figure 1b). Further examination revealed adenoma sebaceum (Figure 1a) and shagreen patch (Figure 1c).

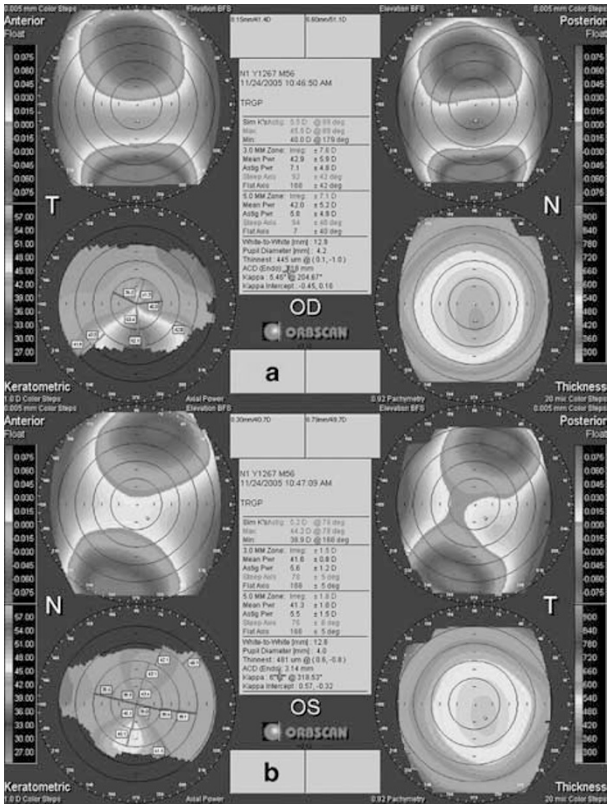

Figure 1 Corneal topography (Orbscan).

Corneal topography (Figure 2) showed corneal changes characteristic of keratoconus with inferior steepening on the right (Figure 2a) and relatively unaffected left eye (Figure 2b).

He is fit and well with no epilepsy, mental illness, renal disease, or atopy. He has no risk factors of keratoconus such as steroid use or vernal conjunctivitis. There is no family history of tuberous sclerosis or keratoconus.

This patient was referred for gas-permeable contact lens.

Tuberous sclerosis or Bourneville's disease is a phakomatoses that can be autosomal dominant or sporadic. Typical ocular findings include retinal and optic nerve astrocytic hamartomas.[1] Less commonly reported associations include cataracts,[2] premacular gliosis,[3] and iridociliary hamartomas.[4] To the best of our knowledge, the association to keratoconus has never been published.

Tuberous sclerosis is caused by mutations in 9q34 (*TSC1*) and 16p13 (*TSC2*), which code for tumour suppressors hamartin and tuberin, respectively. Therefore, their association with hamartomas and various neoplasms[5] is not surprising. However, an association to an ecstatic corneal pathology is difficult to explain.

The pathogenesis of keratoconus is genetically heterogeneous and most commonly sporadic, but various genetic loci have been identified including 5q14.3–q21.1,[6] 3p14–q13,[7] and 16q22.3–q23.1.[8] It is difficult to explain the link on genetic terms as no loci for keratoconus have been found near *TSC1* or *TSC2*. Unlike *PKD1* (polycystic kidney disease), which lies adjacent to *TSC2* at 16p13.3, the loci for keratoconus have been located on chromosomes 5, 3, and 16q. However, studies have shown HLA-A28 to be associated with both tuberous sclerosis[9] and keratoconus.[10] Perhaps, there are genetic linkages between the two diseases but, as there was no family history of either tuberous sclerosis or keratoconus, it may be coincidental as both are known to occur sporadically.

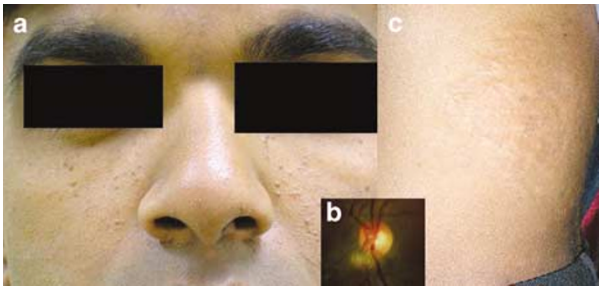

Figure 2 (a) Fundus photograph (hamartoma). (b) Adenoma sebaceum. (c) Shagreen patch.

We did not feel HLA typing was indicated in this patient, as it would not have changed the management of this patient. Furthermore, knowledge of HLA typing would not have enabled us to advise this patient regarding the likelihood of his children developing keratoconus. Therefore, as we still know very little regarding the linkage of the two diseases, the management of this patient included treating the keratoconus and genetic counselling for tuberous sclerosis. Further genetic studies may provide further proof and aid in management of these patients.

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Eye (2007) **21,** 287–288. doi:10.1038/sj.eye.6702536; published online 1 September 2006