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# Lamellar hole-associated epiretinal membrane in retinitis pigmentosa: a surgical correlation

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## Abstract

Retinitis pigmentosa (RP) is a rare hereditary disease, yet it is the commonest cause of retinal dystrophy. Although lamellar hole-associated epiretinal membrane (LHEP) is commonly associated with macular holes, the development of macular holes in RP itself is rare. In this article, we report a rare case of bilateral LHEP in RP, and the surgical outcome of LHEP embedding and internal limiting membrane (ILM) flap in repairing a lamellar macular hole (LMH).

A 57-year-old woman who had RP with bilateral LHEP underwent a combination of cataract and vitrectomy surgery in her left eye. We preserved LHEP tissue and performed an ILM flap to avoid a iatrogenic full-thickness macular hole (FTMH) and facilitate LMH closure. Her right eye was monitored conservatively. At 2 weeks postoperative, the LMH in her left eye was anatomically repaired. There was limited improvement of visual acuity, which could be justified by disruption of the junction between the photoreceptors' inner and outer segments (IS/OS junction) as evidenced by spectral-domain optical coherence tomography (SD-OCT).

The presence of LHEP in LMH is highly associated with disruption of the IS/OS junction and therefore patients should be counselled regarding guarded visual improvement post-vitrectomy. LHEP, which is derived from a Müller cell-driven process, is closely associated with LMH in RP, likely due to the progressive retinal tissue loss as a result from the disease nature of RP. Therefore, we suggest preserving LHEP tissue and performing an ILM flap as an improvised technique to avoid iatrogenic FTMH and facilitate LMH closure.

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## Membran epiretinal yang berkaitan dengan lubang lamela pada retinitis pigmentosa: korelasi pembedahan

### Abstrak

Retinitis pigmentosa (RP) adalah penyakit keturunan yang jarang berlaku, namun ia adalah penyebab distrofi retina yang paling kerap berlaku. Walaupun membran epiretinal yang berkaitan dengan lubang lamela (LHEP) biasanya dikaitkan dengan lubang makula, pembentukan lubang makula pada RP itu sendiri jarang berlaku. Dalam artikel ini, kami melaporkan kes LHEP kedua belah mata yang jarang terjadi dalam RP, dan hasil pembedahan penyisipan LHEP dan membrane pemisah dalaman (ILM) dalam memperbaiki lubang makula lamela (LMH).

Seorang wanita berusia 57 tahun yang menjalani RP dengan LHEP pada kedua belah mata menjalani kombinasi pembedahan katarak dan vitrectomy di mata kirinya. Kami memelihara tisu LHEP dan melakukan flap ILM untuk mengelakkan berlakunya lubang makular ketebalan penuh iatrogenik (FTMH) dan memudahkan penutupan LMH. Mata kanannya dipantau secara konservatif. Pada 2 minggu selepas pembedahan LMH di mata kirinya dibaiki secara anatomi. Peningkatan ketajaman penglihatan adalah terhad, yang disebabkan oleh gangguan persimpangan antara segmen dalaman dan luar fotoreseptor (persimpangan IS / OS) seperti yang dibuktikan oleh tomografi koheren optik-domain spektrum (SD-OCT).

Kehadiran LHEP di LMH sangat berkaitan dengan gangguan pada persimpangan IS / OS dan oleh itu pesakit harus diberi khidmat nasihat mengenai ketidakpastian pada peningkatan visual akuiti selepas jagaan vitrektomi. LHEP, yang berasal dari proses yang didorong oleh sel Müller, berkait rapat dengan LMH dalam RP, mungkin disebabkan oleh kehilangan tisu retina progresif akibat dari sifat penyakit RP. Oleh itu, kami mencadangkan pemeliharaan tisu LHEP dan melakukan flap ILM sebagai teknik improvisasi untuk mengelakkan FTMH iatrogenik dan memudahkan penutupan LMH.

*Kata kunci:* kepek membran dalaman, lubang lamela, membran epiretin, retinitis pigmentosa

## Introduction

Retinitis pigmentosa (RP) is a rare hereditary disease, yet it is the commonest cause of retinal dystrophy. It is characterized by dystrophy of rod and cone photoreceptors, causing decreased night vision and progressive peripheral vision loss, which affect 0.025% of the population worldwide.<sup>1</sup> Fundus examination typically reveals pale optic discs, bony spicule pigmentary changes involving the equatorial retina, arteriolar narrowing, retinal pigment epithelium, atrophy, and cystoid macular oedema.<sup>1</sup>

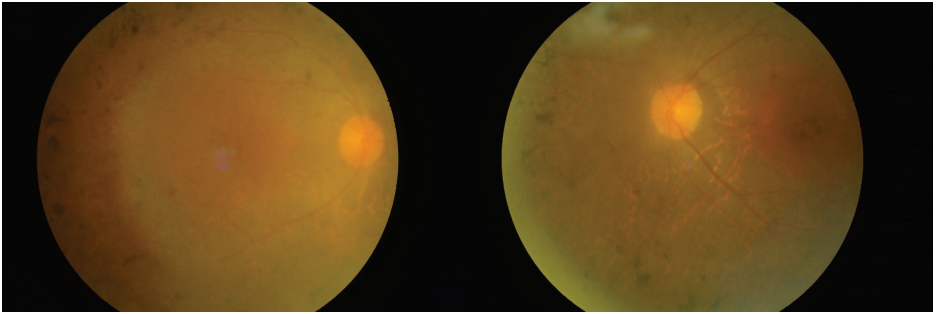
Although lamellar hole-associated epiretinal membrane (LHEP) is commonly associated with macular holes, the development of macular holes in RP itself is rare.<sup>2</sup> LHEP is defined as an homogenous material with medium reflectivity evidenced on spectral-domain optical coherence tomography (SD-OCT), and its presence signifies more severe macular defects and outer retinal disruptions.<sup>2,3</sup> While some studies reported anatomical and functional improvement following vitrectomy surgery for lamellar macular hole (LMH) repair,<sup>4</sup> others suggested conservative management as spontaneous closure is frequently observed and warranted the risk of developing full-thickness macular hole (FTMH) post-vitrectomy.<sup>5,6</sup>

We report a rare case of bilateral LHEP in RP, and the surgical outcome of this patient. The presence of LHEP in LMH is highly associated with disruption of the IS/OS junction and therefore the patient should be counselled regarding guarded visual improvement post-vitrectomy. As a safety measure to prevent iatrogenic FTMH, we propose using LHEP and internal limiting membrane (ILM) flap for LMH repair instead of the conventional LHEP and ILM peeling.

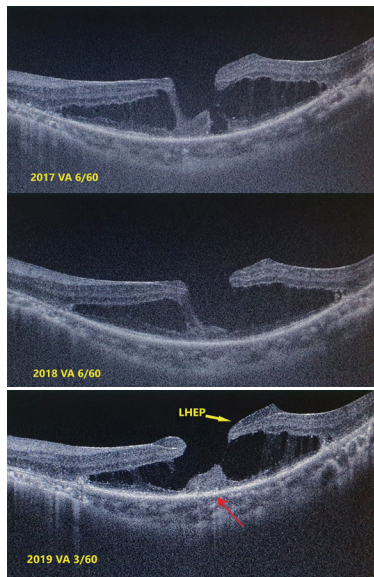
## Case report

A 57-year-old woman who was diagnosed with RP at the age of 20 presented with gradual worsening of vision in both eyes over the past 3 years. On examination, her Snellen best-corrected visual acuity (BCVA) was 6/24 in the right eye and 3/60 in the left eye. Slit-lamp examination of the anterior segment revealed nuclear sclerotic cataract 3+ in both eyes, and posterior segment revealed LMH with LHEP present in both eyes. Narrowing of retinal arterioles and bony spicule pigmentary changes were observed in both eyes. Figure 1 depicts the retinal pathology observed and Figure 2 depicts the serial SD-OCT findings of her left eye.

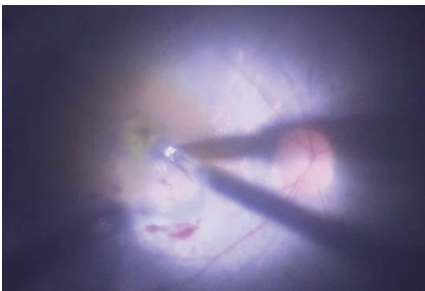
The patient was counselled regarding guarded visual prognosis and risk of developing FTMH, but she decided on going ahead with surgery. She underwent left eye phacoemulsification and implantation of intraocular lens, combined with 23-gauge pars plana vitrectomy. The optic disc gliosis was removed, followed by peeling of the LHEP centripetally using intraocular forceps. The LHEP was left attached to the edge of the LMH, forming a perifoveal crown phenomenon, and



*Fig. 1.* Narrowing of retinal arterioles, bony spicule pigmentary changes and macular hole on the right (*left*) and left (*right*) eye



*Fig. 2.* The serial preoperative SD-OCT images reveal the lamellar hole with LHEP (*yellow arrow*) in the left eye, with evidence of disruption of the IS/OS junction (*red arrow*).



*Video 1.* Steps of lamellar macular hole repair with internal limiting membrane (ILM) flap, starting with optic disc gliosis removal, followed by lamellar hole-associated epiretinal membrane peeling, forming a perifoveal crown phenomenon. Brilliant blue-assisted ILM flap was performed, and the surgery was completed by 20% sulphur hexafluoride gas exchange.

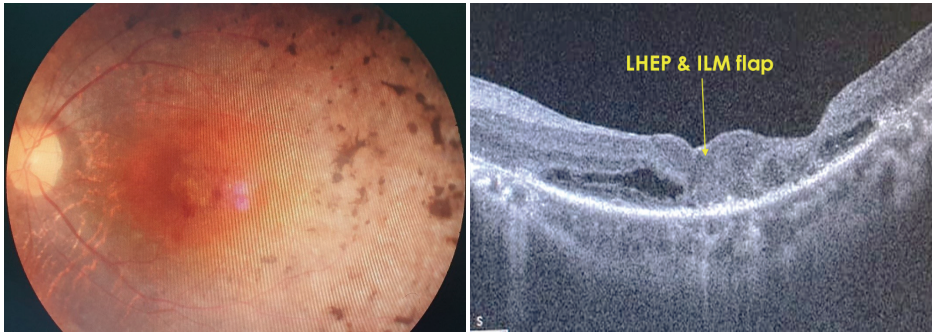


Fig. 3. Fundus photograph (A) and SD-OCT image (B) of the left eye showing repaired lamellar hole using LHEP and ILM flap (yellow arrow) at the 2-week postoperative follow-up.

then gently embedded into the retinal cleavage. We were cautious to minimize unnecessary traction on the fovea. Viscoelastic was injected as protection to the ocular structures, and brilliant blue-assisted ILM flap was performed. The ILM was peeled from the periphery towards the LMH, and then inverted to completely cover the LMH with the embedded LHEP. The surgery was completed by 20% sulphur hexafluoride gas exchange. Video 1 illustrates the steps of the vitrectomy surgery.

At 2 weeks postoperative, the LMH in her left eye was anatomically repaired, but with limited improvement in visual acuity to 6/60. At 2 months postoperative, her BCVA remained stable at 6/60 in the left eye and 6/24 in the right eye. No FTMH development nor macular scarring was observed. Figure 3 describes the anatomically repaired LMH using LHEP and ILM flap in the left eye.

## Discussion

This case report exhibits a rare case of bilateral LHEP in RP, and demonstrates a new technique using LHEP and ILM flap in repairing LMH. Due to the scarcity of LMH formation in RP patients, the mechanism of formation remains unknown.<sup>1</sup> To date, the literature suggests two main mechanisms of LMH formation: tractional LMH due to posterior vitreous detachment with conventional epiretinal membrane (ERM) and intact ellipsoid layer on the one hand, and degenerative LMH due to contraction of LHEP and disrupted ellipsoid layer on the other.<sup>4,5</sup> The latter mechanism is more likely to explain LMH formation in our patient.

Recent studies<sup>3,7</sup> have established that the presence of LHEP is associated with deeper tissue defects, thinner hole bases, and IS/OS junction defects. An immunohistochemical study by Pang *et al.*<sup>7</sup> revealed that LHEP is derived from a Müller cell-driven process originating in the inner retinal layers of the macular defect, which act as a central plug to stabilize the structurally compromised macula.<sup>7</sup> Therefore, progressive retinal tissue loss as a result from the disease nature of RP

may be accountable for the high prevalence of LHEP in RP patients, as observed in our patient.

Vitreotomy for LMH repair in RP remains controversial due to its limited visual improvement postoperatively and the stable nature of LMHs. Pang *et al.*<sup>7</sup> observed that 80% of the LMHs remain functional and morphologically stable without surgical intervention, and spontaneous closure of LMH has been frequently observed in other studies.<sup>2,5</sup> It is not surprising to learn that 20% of the patients with LHEP who underwent vitrectomy developed FTMH,<sup>8</sup> as LHEP is postulated to originate from Müller cell proliferation onto the inner retina.<sup>7</sup> However, there are a growing number of studies demonstrating favourable outcomes in terms of anatomical and functional aspects.<sup>4</sup>

Instead of the conventional vitrectomy, LHEP and ILM peeling, we performed 23-gauge pars plana vitrectomy, LHEP embedding with ILM flap and gas tamponade in our patient. Intraoperatively, LHEP was peeled carefully from the outside to the edge of the LMH, forming a perifoveal crown phenomenon, described by Son *et al.* as a floating, crown-like yellowish tissue with its base attached to the edge of the margin on the macula.<sup>9</sup> We then embedded the LHEP tissue into the LMH to prevent a iatrogenic FTMH, and performed ILM flap to repair the LMH. Shiode *et al.*<sup>10</sup> suggested that performing an ILM flap after LHEP embedding can retain the LHEP in the retinal cleavage during fluid-gas exchange, and that the ILM flap acts as a scaffold to promote the proliferation and migration of glial cells into the LHEP, thus expediting LMH closure<sup>10</sup> and preventing progressive retinal tissue loss.<sup>2</sup>

At 2 weeks postoperative, our patient showed anatomical recovery in the left eye with limited visual acuity improvement, which could be attributed to the preoperative disruption of the IS/OS junction. This outcome is consistent with a study by Ko *et al.*<sup>6</sup> which demonstrated limited postoperative visual acuity improvement for eyes with LHEP despite anatomical improvement given the preoperative disrupted IS/OS line integrity was not restored postoperatively. Furthermore, our patient's right eye, which also had LHEP, remained stable on conservative monitoring. This affirms that LHEP is a stable clinical entity, as established by multiple studies.<sup>6,8</sup>

In this case report we described a rare occurrence of bilateral LHEP in RP, and reported successful anatomical closure of LMH by LHEP embedding and ILM flap technique. However, we were unable to exclude the possibility of recurrence of LHEP, development of secondary ERM, or macular scar formation during long-term follow up, but these were absent at our 2-month post-operative follow up. We hope that this case report could prompt further prospective studies to determine the efficacy and monitor the long-term outcomes of this technique.

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