# Surgical Management of Optic Disc Pit Maculopathy

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**Purpose:** To evaluate the structural and functional outcome of pars plana vitrectomy combined with ILM peel, endo-photocoagulation and gas tamponade for optic disc pit associated maculopathy.

Study Design: Prospective interventional case series

**Place and Duration of Study:** Jinnah Hospital Lahore and Al-Ehsan Eye Hospital Lahore

**Material and Methods:** Seven consecutive patients with unilateral maculopathy associated with optic disc pit underwent pars plana vitrectomy combined with ILM peel, endo-photocoagulation and gas tamponade. Patients were followed up for 6 months after treatment. Main outcomes were determined by optical coherence tomography (OCT) and best – corrected visual acuity (BCVA).

**Results:** Treatment with  $C_3F_8$  tamponade followed by laser photocoagulation in ODP maculopathy patients resulted in resolution of sub-retinal and/or intraretinal fluid in six out of seven patients at the end of six month follow up. The remaining one patient had significant reduction in retinoschisis, as determined by OCT, and fundoscopy, as well as an improvement in anatomical architecture. There was statistically significant improvement in visual acuity in four eyes, remained stable in two eyes and deteriorated in one eye in which we were not able to achieve complete macular reattachment at the end of six months. Central visual field loss after photocoagulation was not clinically appreciable. No post-operative complications of maculopathy occurred during the follow-up period.

**Conclusion:** Given the myriad underlying pathology of sub retinal and intra retinal fluid secondary to optic disc pit associated maculopathy, a complete procedure of pars plana vitrectomy combined with ILM peel, endo-photocoagulation and gas tamponade proved to be an efficient procedure to achieve satisfactory structural and functional out come.

**Key words:** Optic disc pit maculopathy, retinoschisis, photocoagulation, pars plana vitrectomy, internal limiting membrane.

D ptic nerve pits are congenital colobomatous malformation of optic nerve head with a reported incidence of 1 in 11000 patients<sup>1,2</sup>. This condition was first described by Wiethe in 1882<sup>3</sup>. Although some patients remain asymptomatic but maculopathy has been reported in up to 25% – 75% of the patients in different case series and studies<sup>4</sup>. Visual loss secondary to maculopathy involving optic nerve pit is mainly caused by serous macular detachment and retinoschisis. The source and flow of fluid and pathogenesis of maculopathy remain controversial and various clinical mechanisms have been proposed to describe these dilemmas surrounding optic pit maculopathy. One such proposition was given by Lincoff that schisis cavities are initially formed that lead to outer macular hole formation; thus resulting in conduit of fluid from intra-retinal space to sub retinal space<sup>5</sup>. However, Optical coherence tomography

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(OCT) based studies have concluded that there can be a collection of sub retinal fluid in the absence of retinoschisis in cases of optic pit maculopathy<sup>6</sup>. The theory of vitreous origin of fluid has been supported by various previous clinical and histopathology studies like alcain blue staining of vitreous followed by presence of mucopolysaccharides in optic disc pit; videos showing bubbles coming out of the pit after pars plana vitrectomy (PPV) and intraocular gas tamponade; similarly silicone oil has also been described emerging from pit after intraocular tamponade7-9. Cerebrospinal fluid has also been implicated as a potential source of sub retinal fluid and this observation has been substantiated by few observations. MRI contrast dye has been reported to pass into sub retinal space and intraocular silicone oil has been found in intracranial cavity in MRI documented cases10,11. Recent OCT findings have suggested a 3 way connection between intraretinal space, sub retinal space, perineurial space and vitreous cavity; flow and trajectory of fluid may vary among various patients. This hints towards the origin of myriad of surgical options available; all being partially successful and thus the lack of standard surgical approach for optic disc pit maculopathy<sup>12</sup>.

Spontaneous resolution of optic pit maculopathy has been reported in up to 25% of patients but one may encounter macular hole formation, degenerative cystic changes and retinal pigment epithelium degeneration leading to more severe visual loss in the absence of intervention. Therefore, to avoid these sequel observed in the natural course of disease, surgical intervention has been recommended by most investigators<sup>12-14</sup>.

The basic surgical procedure for optic disc pit maculopathy is PPV and intraocular gas tamponade; this procedure has been supplemented with internal limiting membrane (ILM) peel and endolaser photocoagulation to temporal margin of disc<sup>15,16</sup>. Macular buckle, optic nerve sheath fenestration and intra / sub retinal fluid aspiration (recent description) have also been employed as additional procedures with varying functional and anatomic outcomes<sup>17</sup>.

In this study, we report the outcome of 7 cases of optic disc pit maculopathy after PPV, ILM peel, endo laser and intraocular gas tamponade. 2013 to December 2015 after seeking approval from Hospital Ethics Committee and all patients were enrolled in this study after obtaining informed consent. Seven eyes of 7 patients under went standard 23G, 3 port PPV, ILM peel, endolaser and intraocular gas tamponade for optic disc pit associated maculopathy. All surgeries were performed by the same vitreoretinal surgeon. Optic disc pit maculopathy was diagnosed after detailed slit lamp examination, patients complaints, clinical signs and relevant OCT observations. OCT and fundus photographs were taken pre and post operatively.

After standard 3 ports 23G PPV, additional procedures if ILM peel and endo laser were also performed. ILM peel was assisted by Brilliant Peel 0.025% (Fluoron/Geuder, Heidelberg, Germany) dye that preferentially stains ILM without staining cortical vitreous and epiretinal membrane. The nasal extent if ILM peel was till the optic disc margin. Two rows of frequency doubled Nd: YAG laser were applied around the temporal border of optic disc covering the margins of retinal elevation just adjacent to optic disc. Grade I reactions were achieved after applying burns at the level of RPE. After fluid air exchange, a 14% mixture of  $C_3F_8$  and air were used as final endotamponade. All 23G ports were secured using 6 0 Vicryl sutures.

All patients underwent post operative examination at 1st post operative day, 7th post operative day and at 1st, 3rd and 6th post operative month. All patients examined at 1st, 3rd and 6th post operative months were evaluated after documenting their best corrected visual acuity (BCVA) and observations on OCT (Stratus OCT Carl Zeiss Meditec

Inc., Dublin, CA, USA) and (3D OCT – 1 Maestro; Topcon Corporation®, Itabashi, Tokyo, Japan).

SPSS statistical software version 16.0 (SPSS Inc., Chicago, IL, USA) was used for data analysis. Preoperative and post-operative best corrected visual acuity (BCVA) in Log MAR were statistically analysed using Wilcoxon signed rank test. Spearman rank correlation test was used to find relationship between postoperative anatomic and functional results and other relevant parameters. P-value <0.05 was taken as statistically significant.

# MATERIAL AND METHODS

This study was conducted at Jinnah Hospital Lahore and Al-Ehsan Eye Hospital, Lahore from February

# RESULTS

We had 4 male and 3 female patients with mean age of 28.88 +/- 14.55 years (age range 19-66 years). Duration

of loss of vision was between 18 days and 24 months (mean 8.42 +/- 6.03 months. Range of preoperative BCVA was from logMAR 0.3 to 1.30 (mean logMAR 1.00 +/- 0.43). Location wise, 5 pits were located inferotemporally, 1 pit was located surepotemporally and 1 pit was central in location (Fig. 1).

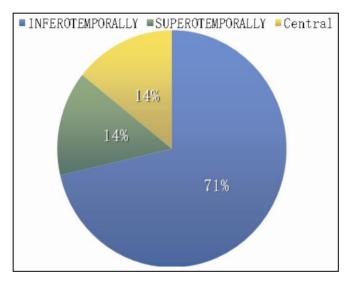


Fig. 1: Location of optic disc pit.

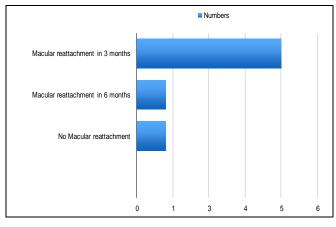
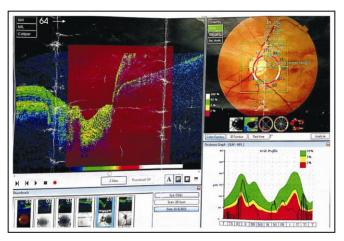


Fig. 2: Time needed before compete macular attachment.

Complete macular reattachment as observed on OCT was achieved in 5 out of 7 eyes in 3 months. One patient achieved macular reattachment at 5th post operative month and 1 patient failed to achieve complete macular reattachment at the end of 6 months follow-up (Figure 2).

By the end of 6 months followup, BCVA improved by at least 2 lines in 4 out of 7 patients. Two patients



**Fig.3a:** Pre-operative OCT and fundus photo of optic disc pit with sensory macular detachment and showing communication between sub retinal space and optic disc pit.

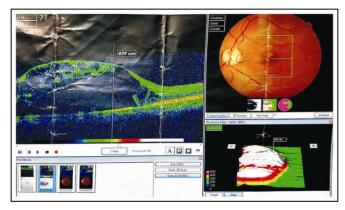
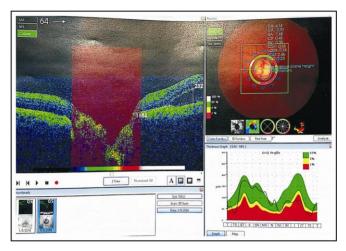


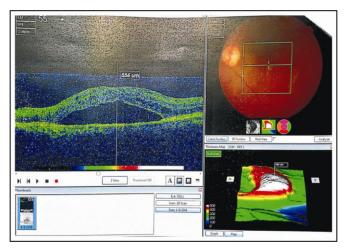
Fig. 3b: Pre-operative OCT and fundus photo of optic disc pit with retinoschisis.

had stable BCVA where as in 1 patient, BCVA deteriorated as compared to her BCVA at the time of presentation. Out of these 2 patients, one patient has an unsuccessful anatomic outcome at the end of 6 months (Figure 3a, 3b, 3c and 3d). Over all, 6 months post operative BCVA ranged from logMAR1. 34 to 0.24 (mean 0.64 +/- 0.54. These finding showed a statistically significant (p-value < 0.005) improvement in BCVA after 6 months of surgical intervention.

Spearman's bivariate correlation concluded that final BCVA was negatively correlated with patients age and duration of symptoms at presentation (P = 0.005, r = 0.6 and P = 0.004, r = 0.63). However, there was no statistically significant correlation between duration of symptoms and pre operative BCVA.



**Fig. 3c:** 6 months post-operative OCT and fundus photo of optic disc pit where macula has failed to reattach completely despite block of communication between optic disc pit and sub retinal space after argon laser photocoagulation.



**Fig.3d:** 6 months post-operative OCT and fundus photo showing complete resolution of retinoschisis but sensory macular elevation persists.

#### DISCUSSION

Optic disc pit presents clinically as a grey round to oval excavation in optic nerve head. Its occurrence is due to lack of perfect closer of embryonic fissure<sup>18</sup>. As our understanding of maculopathy associated with optic disc pit has increased, so have been the options and recommendations for its treatment. Surgeons have tried gas tamponade with or without endolaser barrage, pars plana vitrectomy with or without laser and with or without ILM peeling with varying success rates<sup>19</sup>. Different patterns of maculopathy also influence the anatomical and visual outcome after surgical intervention. One such observation is the presence of multilayer schisis cavities which renders the intervention with poor visual outcome as compared to serous retinal detachment type of maculopathy<sup>20</sup>. Although incomplete understanding of mechanism and management of maculopathy associated with optic disc pit, still 74% of patients with optic disc pit have been found to have certain level of vitreous abnormalities like condensed vitreous strongly attached to pit, vitreomacular traction or incomplete vitreous detachment<sup>21</sup>. Therefore pars plana vitrectomy aided with tamponade, laser or ILM peeling has been widely adopted as a technique to counter this maculopathy with varying success rates.

In our study, we employed the surgical technique of pars plana vitrectomy with ILM peel, endorser and intraocular gas tamponade for treating optic disc pit associated maculopathy. Although if maculopathy is left alone and allowed to follow its natural course, reattachment of macula has been reported but visual outcome is usually very poor<sup>22,23</sup>. Researchers have reported a relatively higher incidence of recurrence after gas tamponade or laser when done alone<sup>24</sup>. Pars plana vitrectomy with or without its adjuncts has been used widely but there are risks associated with pars plana vitrectomy as well (macular hole formation)<sup>13</sup>. In one report, scanning laser ophthalmoscope (SLO) showed a cyst like structure above the pit; and dynamic movement of eyes seemed to exert traction on the pit and may be responsible for development of serous macular detachment associated with optic disc pit<sup>26</sup>. These observations along with above mentioned abnormal vitreous traction role explains why pars plana vitrectomy has become a preferred procedure for optic disc pit maculopathy.

In our study, complete macular attachment was observed in 5 out of 7 patients (72%) at 3 months and 6/7 (86%) at the end of 5 months. Our anatomic results are comparable to Avci et al, who achieved macular reattachment in 11/13 (84%) at the 16 month follow up. Our functional results showed 57% patients had improvement in BCVA of 2 lines or more where as Acvi had such improvement in 84% in his subjects. Just like in our study, he concluded a negative correlation between durations of symptoms and final BCVA<sup>17</sup>. The main difference between the to study groups was that we preferred ILM peeling as an adjunct where as Acvi did his intervention just like us but omitting ILM peeling. Another study showed that 7/8 eyes (87%) achieved retinal reattachment after pars plana vitrectomy and induction of PVD alone without ILM peel, gas tamponade or endophotocoagulation; but complete retinal reattachment in this study was achieved after 1 year as compared to our patients where 6/7 patients achieved complete retinal attachment at end of 6 months.

Although size of macular detachment, presence and severity of retinoschisis and size of optic disc pit may be important factors influencing the therapeutic outcomes of intervention, our study shows that early diagnosis and treatment are fundamental to achieving good functional and anatomic results.

## CONCLUSION

We have presented a study with good functional and structural outcome for optic disc pit associated maculopathy treated by pars plana vitrectomy, endo laser, ILM peel and gas endotamponade. This combination procedure has been effective and we encountered minimal side effects of this procedure. Although, the results cannot be generalised due to limited sample size, further similar studies can further support the therapeutic approach we adopted for this pathology. Despite the difficulty in conducting randomised clinical trial for optic disc pit maculopathy because of its very low incidence, we still need further scientific evidence to tailor our approach in finding the best procedure to address this pathology completely.

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#### **Role of Authors**

Dr. Haroon Tayyab Surgeon and study design. Dr. Tehmina Jahangir Patient evaluation and literature search.

Dr. Akhwand Abdul Najeed Jawad Statistical analysis and discussion writing.

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