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Vitrectomy for macular detachment associated with optic nerve pits *

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Abstract

Objective. To describe the pathogenic mechanism for a central retinal detachment in eyes with a pit of the optic disc. *Design.* Three patients with macular detachment associated with optic nerve head pits were prospectively controlled following pars plana vitrectomy. *Methods.* Three cases of macular detachment associated with congenital optic nerve pit were treated with pars plana vitrectomy and air-fluid exchange and sulphur hexafloride (SF6) gas injection. During vitrectomy posterior hyaloid removal caused visible traction on the elevated retina, when the hyaloid detached. Removing the posterior hyaloid resulted in reattachment of the macular and an increase in the visual acuity without laser treatment. *Conclusion.* We recommend the removal of the posterior hyaloid face during pars plana vitrectomy in patients with central macular detachment associated with a pit of the optic disc.

Introduction

Posterior retinal detachment is a common observation in eyes with pits in the optic nerve head [1–3]. Controversy exists over the source of the subretinal fluid. Liquefied vitreous [4], cerebrospinal fluid [5] or fluids from orbital tissues [6] have been suggested. As the basic pathogenic principle, slit-like defects of the dysplastic herniated retina in the pit of the optic disc are proposed to enable a free communication to the subretinal space. Attempts to repair the macular detachment and thereby to improve the visual prognosis have included binocular patches and bed rest [7, 8], ACTH and corticosteroid [9] and mannitol [10] treatment. Furthermore surgical intervention using diathermy [11] or scleral folding [12] has been proposed. At present photocoagulation [13–18], pneumatic retinopexy [19, 20] and vitrectomy [7, 8, 21, 22] are recommended.

We present three patients with macular detachment due to a congenital pit of the optic disc. They were successfully treated with vitrectomy and gas endotamponade. During vitrectomy surgery we identified and removed the posterior hyaloid face from the retinal surface in all eyes. No laser treatment was performed.

Report of cases

Case 1

A 16-year-old girl was admitted in March 1993 with diminished visual acuity of 2-month duration relating to a temporal optic pit in her left eye. The visual acuity was reduced to 20/200. Ophthalmoscopic examination revealed an opaque central elevation of the retina. Biomicroscopy showed an oval hole at the macula in the outer layer of the retina. Posterior vitreous detachment, even localised, could not be observed. Fluorescein angiography demonstrated hypofluorescence of the pit area in the arterial and recirculation phase. Pigment epithelial alterations were present, corresponding to the area of the macular detachment. Slight leakage of fluorescein into the subretinal space was noted on the disc margin at the edge of the pit and in the centre of the detached retina. In the late phase of the angiogram the optic pit became hyperfluorescent (Figure 1, Inset).

The patient was scheduled for pars plana vitrectomy 6 weeks later, to await a spontaneous resolution of the macular detachment. On examination after this period, the visual acuity in her left eye remained 20/200. The fundus examination revealed an unchanged macular detachment.

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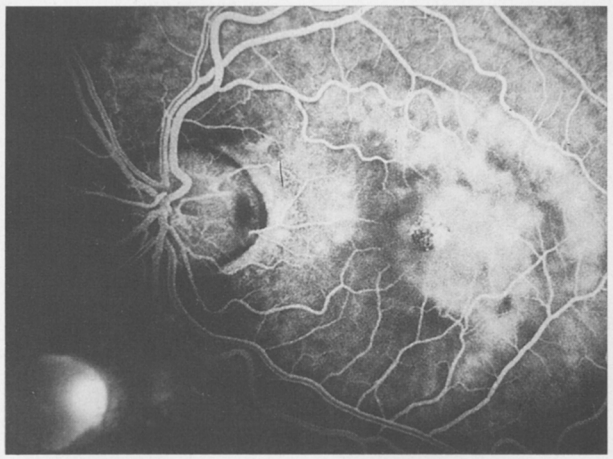

Figure 1. Case 1, fluorescein angiography. Recirculation phase. Note the hypofluorescent pit, the outer layer macular hole and the diffuse leakage under the macular region. Inset: Hyperfluorescence of the optic pit in the late phase of the angiogram.

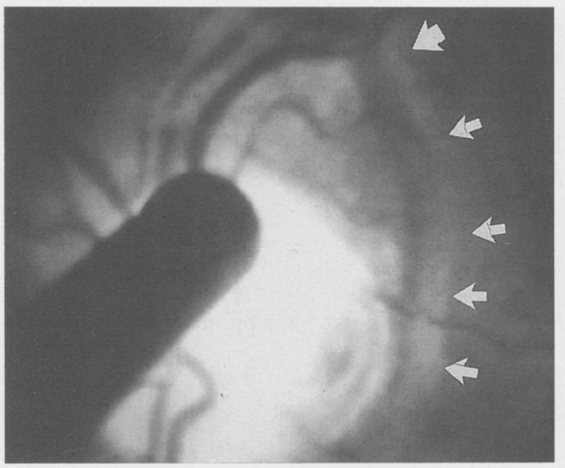

Figure 2. Case 1, operation. Removing the posterior hyaloid caused visible traction (undulation, arrows) on the elevated retina, when the hyaloid detached.

During surgery, a complete pars plana vitrectomy was performed. The face of the posterior hyaloid was identified near the optic disc using suction with an extrusion cannula. Posterior hyaloid removal caused visible traction, undulating the central detached retina (Figure 2, arrows) when the hyaloid detached. The operation was ended by gas-fluid-exchange using a 20% non-expandable mixture of sulphur hexafluoride (SF6). No laser treatment was performed.

On follow-up examination one month later, the visual acuity improved to 20/50. The macula was attached, and with fluorescein angiography no leakage into the subretinal space could be verified. At the 6-month postoperative follow-up examination visual acuity improved to 20/32 and the macula showed no subretinal fluid (Figure 3). At the 12-month follow-up examination her visual acuity had further improved to 20/25, which remained stable at the 2- and 3-year follow-up intervals.

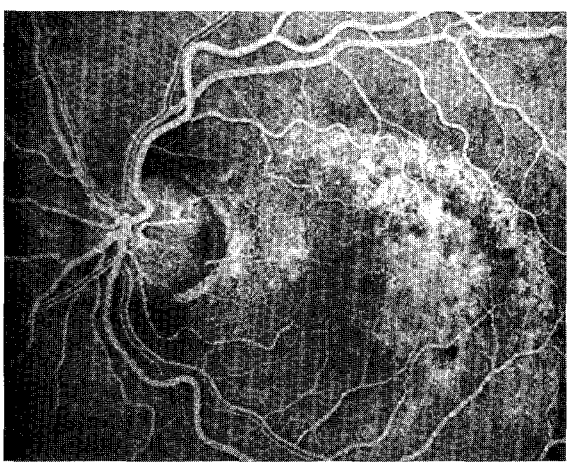

Figure 3. Case 1, fluorescein angiography at the 6-month follow-up. Recirculation phase. Fluorescein angiography showed attached macular and mottled retinal pigment epithelium. Leakage of fluorescein into the subretinal space could not be verified.

Case 2

A 16-year-old boy was admitted in December 1992 with a history of a submacular detachment of four-day duration in his left eye. Visual acuity was 20/50 in the affected eye. Ophthalmoscopy revealed in tiny inferotemporal optic pit combined with an inferior coloboma of the choroid. Biomicroscopy showed a target-like elevation of the fovea with a cystic centre. Awaiting a spontaneous resolution the patient was observed three and six months later. When he returned in June 1993, the visual acuity in his left eye deteriorated to 20/100. Fundus examination revealed serous macular detachment as before.

Pars plana vitrectomy without photocoagulation treatment, peeling of the posterior hyaloid, gas-fluid exchange (20% – SF6) and face-down reattached position the macula.

At 4 weeks, the visual acuity was 20/60 and the central retina was flat. At 6 months the visual acuity

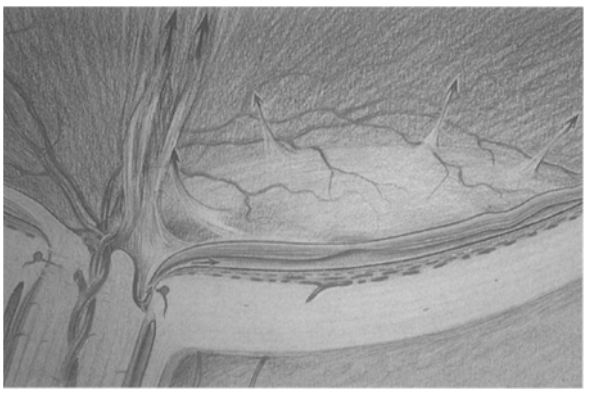

Figure 4. Schematic illustration of the pathogenesis of macular detachment complicating an optic pit. Vitreous traction on the macular region results in the leakage of fluid from the vitreous cavity toward the subretinal space.

improved to 20/40 and remained constant at 12 months after surgery.

Case 3

A 14-year-old boy was admitted in June 1991 with diminished visual acuity relating to a temporal optic pit of his right eye. The visual acuity was reduced to 20/30. Ophthalmoscopic examination revealed a central elevation of the retina and the pigment epithelium beneath it had become mottled. At first no treatment was given to this patient. Six months later no changes were found in his right fundus, but visual acuity decreased to 20/40. The patient was treated with 28 spots of argon green laser photocoagulation in 2 rows along the temporal disc margin. When he was seen 1 month later the visual acuity had improved to 20/30 but the subretinal fluid did not resolved completely. Visual acuity remained constant at 20/30 until May 1994. Then he noted a deterioration in vision. At examination the visual acuity was 20/50 and biomicroscopy revealed a slight increase in the amount of subretinal fluid.

Pars plana vitrectomy, without photocoagulation treatment, by removing the posterior hyaloid and gas-fluid exchange was performed. The retina was flat postoperatively and visual acuity in this eye improved to 20/25 over the next 6 weeks. At this examination we revealed a flat peripheral detachment in the inferior region, caused by dialysis of the ora serrata. Therefore we paced an encircling band in a secondary procedure. One month later the visual acuity was again 20/25.

Discussion

Deterioration of visual acuity (Table 2) in patients with congenital pit of the optic nerve head usually occurs as a result of complications of chronic serous detachment of the retina, macular pigmentary alterations and macular hole formation. Spontaneous resolution of the retinal detachment associated with optic pits is well known and occurs in approximately 31% of cases (Table 1). However, the presence of subretinal fluid for more than 1 year causes an irregular damage to the pigment epithelium of the affected area [23]. For this reason in our cases we awaited a spontaneous reattachment of the retina, within a limited time interval to the onset of the symptomatic macular detachment, before we performed pars plana vitrectomy.

According to the mechanical concept of Sugar [4], who suggested that the macular detachment in eyes with congenital optic pits is due to a leakage of aqueous from the vitreous chamber through the pit into the subretinal space, the first treatments using xenon arc photocoagulation were performed by Gass [5] and Meyer-Schwickerath [12] in 1966. Their goal was to create a chorioretinal adhesion at edge of the optic disc to resolve the submacular fluid. Jack [24] in 1969 reported the first successful photocoagulation of this condition. Nevertheless, macular reattachment could be achieved using photocoagulation in only 48% of cases (Table 1). When laser treatment is carried out, krypton red laser is recommended. Krypton laser offers the theoretical advantage of a burn in the outer retina and choroid, thus treatment might therefore less likely to damage to the nerve fibre layer [7, 8, 18]. In contrast to the findings reported by Brown et al. [3], who found that posterior vitreous detachment was clinically apparent in 80% of eyes with serous macular detachment, we found that all eyes in our series showed no clinical evidence of posterior vitreous detachment. Our findings correlate with the data by Gordon et al. [25], Gass [5], Bonnet [19] and Akiba et al. [26].

Zimmermann, quoted by Ferry [27], thought that traction exerted upon the macula is probably the mechanism responsible for the ultimate detachment of the macula. Such traction might result from the lengthened, indirect course which must be followed by ganglion cell axons arising from part of the retina associated with the optic pit, and from changes in the glial tissue in the pit itself.

Akiba et al. [26] postulated that a condensed vitreous strand that extend to the optic pit as described in several studies may represent the anomalous Clo-

Table 1. Anatomic results of macular detachment operations in eyes with optic pit vs. spontaneous follow-up

Study	No.		No treatment		Photocoagulation		PR ± Laser		PPV/Gas ± Laser	
	R	E	S	U	S	U	S	U	S	U
1 Alexander et al. [21]	2	2	–	–	–	–	–	–	2	0
2 Annesly et al. [18]	5	5	–	–	4	1	–	–	1	0
3 Bonnet et al. [19]	25	20	2	0	1	9	7	6	–	–
4 Brockhurst [15]	6	6	–	–	6	0	–	–	–	–
5 Brown et al. [3]	20	20	5	15	–	–	–	–	–	–
6 Cox et al. [7]	14	14	–	–	1	2	–	–	10	1
7 Gass [5]	5	4	1	1	0	2	–	–	–	–
8 Jack [24]	1	1	–	–	1	0	–	–	–	–
9 Kottow [16]	2	2	–	–	2	0	–	–	–	–
10 Kwang et al. [22]	1	1	–	–	–	–	–	–	1	0
11 Lincoff et al. [20]	9	9	–	–	0	3	3	2	3	1
12 Lincoff et al. [28]	15	14	0	4	1	6	–	–	2	1
13 Mustonen et al. [13]	3	3	–	–	3	0	–	–	–	–
14 Rubinstein et al. [32]	16	14	1	13	–	–	–	–	–	–
15 Schatz et al. [8]	6	5	–	–	2	1	–	–	3	0
16 Sobol et al. [35]	15	15	4	11	–	–	–	–	–	–
17 Snead et al. [36]	1	1	–	–	–	–	–	–	1	0
18 Sugar [33]	12	11	10	1	–	–	–	–	–	–
19 Sugar [34]	1	1	1	0	–	–	–	–	–	–
20 Sugar [4]	6	3	0	3	–	–	–	–	–	–
21 Theodossiadis [17]	14	14	0	8	4	2	–	–	–	–
22 Tillmann et al. [14]	1	1	–	–	1	0	–	–	–	–
23 Vogel et al. [12]	2	2	–	–	0	2	–	–	–	–
Total	182	168	24	54	26	28	10	8	23	3
Success [%]			31		48		56		88	

Gas – air: study 6, 12, 14; SF6: study 1, 3, 6, 10, 12; perfluorocarbon gases: study 3, 11, 12. No treatment – means no surgical treatment. No. – number of reported eyes = R, and number of evaluated eyes = E. Photocoagulation – Xenon: study 4, 7, 8, 13, 18, 19, 20; Argon: study 1, 3, 4, 6, 9, 10, 11, 12, 14, 18; Krypton: study 2, 6. PPV/Gas ± Laser – pars plana vitrectomy, fluid/gas- or fluid/air-exchange, with or without laser treatment and after unsuccessful photocoagulation or pneumatic retinopexy. PR ± Laser – Pneumatic retinopexy, with or without laser treatment, and after unsuccessful photocoagulation; S = successful treated; U = unsuccessful treated.

quet's canal. He believed that eyes with an optic pit have a congenital malformation of the vitreous body combined with an anomaly of the optic nerve head. Vitreous traction to the macula was suggested as having an important role in developing macular detachment. The condition of the vitreous toward the optic disc and the macular region might be the reason why schisis-like separations of the internal layers of the retina over the area of laser-induced chorioretinal adhesions, as observed by Lincoff et al. [28], would be likely to result in a macular redetachment. This could probably be the reason why laser treatment failed in Case 3 of our series.

Bonnet [19] and Lincoff et al. [20] reported successful treatment of macular detachment associated with optic nerve pits by pneumatic retinopexy in 56% of cases (Table 1). Bonnet [19] in her series of eyes treated with gas injection observed lacunae in the posterior vitreous gel as well as a fibrous appearance of the posterior vitreous cortex. She interpreted these changes as representing a posterior vitreous separation induced by gas. The intention of Lincoff et al. [20] has been to displace an outer layer detachment rather than to compress the retina at the edge of the disc to enhance the effect of laser treatment. They emphasised that a 40% filling of gas combined with a 30 degree gaze-down position is optimal for displacement of the central retinal elevation in the optic pit syndrome. Instead of laser application to edge of the optic disc, in one of his cases he applied a horizontal line of burns along the inferior

Table 2. Visual acuity from 64 patients with congenital optic pits and associated macular detachment who do not undergo surgical treatment

Visual acuity	Number of eyes at time of initial detachment		Number of eyes at most recent examination	
	n	%	n	%
20/ 16 – 20/ 32	20 (8)	31 (40)	13 (5)	20 (25)
20/ 40 – 20/ 80	26 (8)	41 (40)	16 (4)	25 (20)
20/100 – 20/200	9 (1)	14 (1)	11 (5)	17 (25)
20/400 – CF	9 (3)	14 (15)	24 (6)	38 (30)

Data from Brown [3], Sobol [35], Sugar [4, 33, 34], Gass [5], Theodossiadis [17] and Lincoff [28]. Data from Bonnet [19] and Rubinstein et al. [32] had to be excluded, because visual acuity of the initial or the most recent examinations was missing. Data from Brown's study alone [3] were put in parentheses. In Brown's [3] study mean follow-up was 4.5 years, for the rest of the group mean follow-up was 5.5 years. CF – counting finger.

Table 3. Visual results from photocoagulation in 38 patients with congenital optic pit and associated macular detachment

Visual acuity	Number of eyes at time of initial detachment		Number of eyes at most recent examination	
	n	%	n	%
20/ 16 – 20/ 32	2	5	10	26
20/ 40 – 20/ 80	6	16	16	42
20/100 – 20/200	21	55	7	19
20/400 – CF	9	24	5	13

Data from Gass [5], Cox et al. [7], Schatz et al. [8], Vogel et al. [12], Mustonen et al. [13], Tillmann et al. [14], Brockhurst [15], Kottow [16], Theodossiadis [17], Annesly et al. [18], Jack [24] and Lincoff et al. [28]. Data of 6 eyes from Bonnet [19] had to be excluded, because visual acuity of the initial examinations was missing. Data of 3 other eyes from Bonnet [19] had to be excluded, because pneumatic retinopexy had been performed in follow-up. Data of 1 eye from Cox et al. [7], 1 eye from Schatz et al. [8], 1 eye from Annesly et al. [18], 3 eyes from Lincoff et al. [20] and 1 eye from Lincoff et al. [28] had to be excluded because pars plana vitrectomy had been performed in follow-up. Mean follow-up for the group was 1.8 years. CF – counting finger.

arcade to confine the pneumatic displacement of the outer layer detachment below.

Inspired by Schatz in 1982, Alexander et al. [21] successfully performed vitrectomy and gas-fluid exchange combined with photocoagulation for reabsorption of the subretinal fluid in eyes with macular detachment associated with optic pits in 2 cases. McDonald [29] indicated that a determined effort to identify and remove the posterior hyaloid face from

Table 4. Visual results from pneumatic retinopexy in 5 patients with congenital optic pit and associated macular detachment

Visual acuity	Number of eyes at time of initial detachment		Number of eyes at most recent examination	
	n	%	n	%
20/ 16 – 20/ 32	0	0	1 (4)	20 (31)
20/ 40 – 20/ 80	2	40	3 (4)	60 (31)
20/100 – 20/200	2	40	0 (4)	0 (31)
20/400 – CF	1	20	1 (1)	20 (7)

Data from Lincoff et al. [20]. Data of the final visual acuity of 13 eyes from Bonnet [19] were put in parentheses. Mean follow-up for both groups was 1.9 years. CF – counting finger.

Table 5. Visual results from pars plana vitrectomy in 26 patients with congenital optic pit and associated macular detachment

Visual acuity	Number of eyes at time of initial detachment		Number of eyes at most recent examination	
	n	%	n	%
20/ 16 – 20/ 32	0	0	9	35
20/ 40 – 20/ 80	4	15	6	23
20/100 – 20/200	13	50	8	30
20/400 – CF	9	35	3	12

Data from Cox et al. [7], Schatz et al. [8], Annesly et al. [18], Lincoff et al. [20], Alexander et al. [21], Kwang et al. [22], Snead et al. [36] and Lincoff et al. [28]. Mean follow-up for the group was 2.2 years. CF – counting finger.

the retinal surface is appropriate. Vitrectomy has been used with considerable success to treat sensory macular detachment associated with optic pit. To our knowledge, results of 25 eyes are reported in literature to date (Table 1). Reattachment could be achieved in 88%. Comparing visual results of photocoagulation, pneumatic retinopexy and pars plana vitrectomy the latter technique shows the most advantage (Tables 3–5).

In our opinion the combination of traction on the retina in the presence of a retinal hole is the cause of rhegmatogenous retinal detachment. Strong suction forces of the pigment epithelium and the choroid counteract such a development [30]. Traction transferred by a congenital malformation of the vitreous body [26] to the macular, as observed during removal of the posterior hyaloid in our series, in the presence of a channel for accumulating extracellular fluids from liquefied vitreous [4] or the subarachnoidal space [31] might be the reason for the retinal detachment in optic pit syndrome (Figure 4). Since the fistula in the dysplastic retina of

the pit of the optic disc is small and leakage to the subretinal space is compensated by strong suction forces, the detachment might be limited. The configuration of retinal detachment might depend on the extension of the adherence of the posterior hyaloid to the central retina.

To date we could not determine if we would achieve the same anatomical results only by core vitrectomy without removing the posterior hyaloid. This would support the hypothesis of Bonnet [19], who described the macular detachment in optic nerve head pits by traction of the vitreous on the roof of the optic pit allowing the vitreous liquid to reach the subretinal space. However, our observations during vitrectomy support the assumption, that the presence of an attached posterior hyaloid is of great importance. Final conclusion could only be achieved by controlled studies. With regard to our own results and to the results reported in the literature, we suppose that anterior/posterior plus tangential traction, similar to eyes with macular hole formation [37], is the essential part in the development of the macular detachment. Alterations of the retinal pigment epithelium are secondary changes.

Removing the posterior hyaloid from the macula during vitrectomy requires experienced vitreoretinal surgery, and subsequently a reattachment of the macula could be achieved without producing chorioretinal adhesions at the edge of the optic pit and without drainage of the subretinal fluid by retinotomies. Probably gas-fluid exchange at the end of surgery as performed in our series might be unnecessary.

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