Definition, history and synonyms

There are two definitions of hypotony. ‘Statistical’ hypotony can be defined as an intraocular pressure (IOP) less than 6.5 mmHg, which corresponds to more than three standard deviations below the mean; ‘clinically significant’ hypotony represents the condition where the IOP is low enough to result in visual loss (Pederson 1996). There are several causes of visual loss secondary to low IOPs, including hypotony maculopathy, keratopathy, cataract formation, choroidal effusion, optic nerve oedema, irregular astigmatism and even phthisis bulbi (Fannin et al. 2003). This review will explore the definition, mechanisms, clinical findings and treatment of hypotony maculopathy.

Hypotony maculopathy was first described by Dellaporta (1954). In this publication, he assembled four cases that had been reported between 1946 and 1954 (Renard 1946; Dellaporta 1948; Pau 1950). He recognized that the condition occurred usually after antiglaucomatous surgery or after perforating eye injuries, and stressed that ‘the striking common characteristic of the four reported eyes was ocular hypotony of about 8–10 mmHg’. The syndrome was characterized by hypotony associated with fundus abnormalities, which included papilloedema, vascular tortuosity and chorioretinal folds. Since the papilloedema was usually of moderate degree and was not associated with retinal haemorrhages, the term ‘papilloedema _ex vacuo_’ was proposed in order to clearly distinguish this condition from the true papilloedema because of increased intracranial pressure (Dellaporta 1954).

Several years later, Gass (1972) coined the term ‘hypotony maculopathy’, chosen only to emphasize that alterations in the macular region were primarily responsible for the loss of central vision. Although ‘hypotony chorioretinopathy’ might be a more accurate term to describe the fundoscopic changes, which may involve virtually the entire ocular fundus, we decided to maintain the term ‘hypotony maculopathy’ throughout this
Clinical findings

Hypotony maculopathy corresponds to a condition where hypotony leads to papilloedema associated with wrinkling or folding of the retina and choroid throughout the posterior pole. In the macular area, there are several fine retinal folds radiating outward from the fovea (Fig. 1). The folding results in distortion of the neurosensory elements of the retina and reduction of the anteroposterior diameter of the eye, leading to a relative hyperopia. The chorioretinal folds are particularly evident in the macula. In this area, the very thick perifoveal retina surrounding the very thin foveal retina is thrown into radial folds around the fovea (Gass 1972).

With the onset of hypotony, the posterior scleral wall probably begins to shrink or collapse, throwing the choroid and retina into folds. As the wrinkling or folding becomes more prominent, the relative compression of pigment epithelial cells in the trough of the fold and the relative thinning of the pigment epithelium over the crest produce the alternate dark and light coloured streaks that can be observed ophthalmoscopically (Fig. 2). In prolonged hypotony, the intensification of the alternate light and dark streaks in the fundus is probably caused by alterations in the structure and pigment content of the retinal pigment epithelial cells (Figs 2 and 3).

Acutely acquired chorioretinal folds usually produce visual dysfunction caused by distortion of the overlying retinal receptors. In hypotony maculopathy, the distortion of the photoreceptors is probably associated with other hypotony-induced changes (such as irregular astigmatism) that exacerbate the visual disturbance caused by the chorioretinal folds. These folds may have a horizontal, oblique or vertical orientation, although an irregular or radiating pattern may be present. The longer the duration of the folds, the more prominent they appear.

Cystoid macular oedema caused by abnormal retinal capillary permeability may ensue secondary to a reduction in the interstitial pressure (Kokame et al. 2001; Stefansson 2006). However, this is not a common finding, and – even if present – is of secondary importance as a cause of visual loss in these eyes. The retinal vessels appear tortuous, and the retinal veins engorged (Figs 1 and 3). Serous detachment of the peripheral choroid is usually absent at indirect ophthalmoscopy (Gass 1972), but may be recognized with ultrasound biomicroscopy (Coleman 1995).

Papilloedema is probably the result of anterior bowing of the lamina cribrosa, constricting axonal bundles in the lamina scleralis and reducing orthograde and retrograde axoplasmic transport (Minckler & Bunt 1977) (Figs 1 and 3). Disc swelling is less frequent in patients with advanced optic nerve damage, because there are fewer axons left to suffer changes in axoplasmic flow.
Imaging

Fluorescein angiography is helpful in demonstrating the chorioretinal folds, which in relatively mild degrees may be overlooked (Fig. 4). It is also useful in differentiating folds of the choroid and retinal pigment epithelium (RPE) from folds in the retina, which do not alter background fluorescence. Early in the course of hypotony, fluorescein angiography shows an irregular increase in background choroidal fluorescence corresponding to the crest of the choroidal folds and also shows some evidence of leakage of dye from the capillaries on the optic nerve head, but usually not from the retinal capillaries (Gass 1972). Characteristic changes in the background choroidal fluorescence are caused by folding of the choroid and RPE. Intensification of the choroidal fluorescence occurs along the crest of the choroid and RPE and produces a series of relatively hyperfluorescent streaks that are evident as early as the arterial phase. The hyperfluorescent streaks are caused by the relative thinness of the RPE on the crest, the greater thickness of the pool of choroidal dye beneath the crest and the shorter course of the incident blue and reflected yellow-green light through the RPE on the crest. The troughs of the folds appear relatively hypofluorescent. This results in angiographic finding of narrow dark lines running within a background of normal or slightly intensified background choroidal fluorescence.

Indocyanine green angiography may reveal multiple hypofluorescent spots in many parts of the fundus, sector hypofluorescent areas, dilatation and tortuosity of the choroidal vessels in the posterior pole of hypotonic eyes, even if fluorescein angiography is not able to detect them (Masaoka et al. 2000).

B-scan ultrasonography is especially useful when the fundus is not easily visualized. It can help in excluding the presence of ciliochoroidal detachment, suprachoroidal haemorrhage and retinal detachment, which are not commonly seen in cases of hypotony maculopathy. Ultrasonography usually demonstrates some flattening and thickening of the posterior sclera and choroid, but identifying the chorioretinal folds may be difficult (Cappaert et al. 1977) (Fig. 5).

Ultrasonic biomicroscopy can be employed (Fig. 6) to further evaluate the anterior chamber depth, the position of the ciliary body and the presence of anterior ciliary detachment (Roters et al. 2002). Furthermore, it can be used to identify a cyclodialysis cleft, one of the important causes of hypotony maculopathy (Blende et al. 1999; Chan et al. 2000). Intraoperatively, the ciliary body can be directly visualized to evaluate rotation and traction using endoscopy (Hammer & Grizzard 2003; Gnanaraj et al. 2005).

Optical coherence tomography (OCT) of the posterior pole can help to better demonstrate subtle macular fluid or folds. OCT can be helpful in
diagnosing suspected hypotony maculopathy in patients with reduced visual acuity (VA) and normal ocular examination, except for low IOPs. Budenz et al. (2005) suggested that because retinal folds are typically oriented in the 0–180° axis, careful review of all radial line scans may be necessary to diagnose this condition (Fig. 7). Martinez de la Casa et al. (2003) reported the case of a 40-year-old man who underwent trabeculectomy and showed hypotony and VA loss.

Aetiology, histopathology and mechanisms

Any condition leading to reduced IOP may result in hypotony maculopathy. Causes of hypotony are listed in Table 1, although the discussion of these extends beyond the scope of this article.

Hypotony occurs when aqueous humour production does not keep pace with outflow. Outflow may be greater than usual, as seen with a wound leak, an overfiltering bleb or a cyclodialysis cleft. Conditions that alter ciliary body function, such as iridocyclitis, ciliochoroidal detachment or hypoperfusion, may cause inadequate aqueous humour production (Newhouse & Beyrer 1982; Toris & Pederson 1987; Pederson 1996; Kato et al. 1999; Fannin et al. 2003). Although hypotony maculopathy is usually seen after glaucoma filtering surgery, especially with adjunctive mitomycin C (MMC), it can also be observed following other ocular conditions (Hatton et al. 1998; Foster et al. 1999; Deramo et al. 2001; Ichibe et al. 2002).

Inflammation may play a key role in the evolution of hypotony. It causes increased permeability of the blood–aqueous barrier. Choroidal fluid is believed to accumulate as a result of enhanced uveoscleral outflow and decreased aqueous humour production, a cycle that is often perpetuated once choroidal effusion develops. A ring of anterior choroidal fluid can rotate the ciliary body forward, impairing its ability to produce aqueous humour (Weekers & Delmarcelle 1953; de Smet et al. 2005).

Dellaporta (1954) believed that, in hypotonic eyes, the protruding nerve head pulled the nerve-fibre layer causing friction between the retina and the pigment epithelium of the retina along the nerve fibres. This friction would cause irritation and subsequent gradual increased pigmentation. He also believed that the tendency for the choroidal folds to have a linear branching course radiating in a temporal
direction away from the optic disc, and to have a cobbledstone, flagstone, hexagonal or quilt-like pattern nasally, was probably caused by several factors including structural differences between the choroid and sclera and unequal forces exerted in the collapse of the ocular coats during eye movement by virtue of the oblique and nasal insertion of the optic nerve head.

Gass (1972) proposed the theory that is now accepted to explain the mechanism of hypotony maculopathy. He suggested that hypotony could cause the scleral wall to collapse inward, resulting in redundancy of the choroid and retina, which would lead to chorioretinal wrinkling. The unusual configuration of retinal folding is a result of the peculiar anatomy of the retina in the macula. As the antero-posterior diameter of the vitreous cavity decreases in hypotony, the very thick perifoveal retina surrounding the very thin foveal retina is thrown into radial folds around the fovea (Gass 1972) (Fig. 8).

Several authors have subsequently confirmed that any condition causing a reduction in the area of the inner surface of the sclera (scleral thickening or scleral shrinkage) may cause the inner portion of the choroids, the overlying retinal pigment epithelium and the outer retinal layers to be thrown into a series of folds or wrinkles (Dellaporta 1950; Norton 1969; Hyvarinen & Walsh 1970; Kroll & Norton 1970; François & DeLaey 1971; Von Winning 1972; Newell 1973; Cangemi et al. 1978; Kalina & Mills 1980; Morris & Sanders 1980; Lebuisson et al. 1981; Gass 1987).

The origin of papilloedema was explained in a series of experimental studies. According to Minckler & Bunt (1977), hypotony could lead to anterior bowing of the lamina cribrosa, constricting axonal bundles in the lamina scleralis and reducing orthograde and retrograde axoplasmic transport. In a subsequent study, Floyd & Minckler (1983) demonstrated that leakage from the choriocapillaris could be an additional source of the fluid that accumulates in the disc. They speculated that swollen axons, caused by blockage of the axoplasmic transport, could compromise the blood flow to the optic nerve, resulting in hypoxia, endothelial cell damage and leakage. Histopathologic findings in eyes with hypotony have been described by Collins (1917), who demonstrated thickening and folding of the sclera and folding of Bruch’s membrane, associated with accumulation of retinal pigment epithelium in the depths of the choroidal folds. Histopathology of hypotonic eyes may also demonstrate generalized oedema of the uvea, retina and optic nerve, as well as a proteinaceous fluid in the suprachoroidal space (Volcker & Naumann 1979).

Incidence and risk factors

The initial literature on hypotony maculopathy included several reports of cases; these did not allow the evaluation of its incidence. More recently, hypotony maculopathy has been reported to occur in up to 20% of cases of glaucoma filtering surgery (Whiteside-Michel et al. 1992; Costa et al. 1993c; Kitazawa et al. 1993; Table 1.

<table>
<thead>
<tr>
<th>Causes of hypotony</th>
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<tbody>
<tr>
<td>Postoperative hypotony</td>
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<tr>
<td>Hypotony after trauma</td>
</tr>
<tr>
<td>Bilateral hypotony</td>
</tr>
<tr>
<td>Miscellaneous forms of hypotony</td>
</tr>
<tr>
<td>Wound leak</td>
</tr>
<tr>
<td>Overfiltration</td>
</tr>
<tr>
<td>Iridocyclitis</td>
</tr>
<tr>
<td>Ciliochoroidal detachment</td>
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<tr>
<td>Retinal detachment</td>
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<tr>
<td>Cycloidalysis</td>
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<tr>
<td>Sceral perforation</td>
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<td>MMC-induced toxicity of the ciliary body</td>
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</table>

MMC, mitomycin C.
Although the description of hypotony maculopathy dates from 1955 (Dellaporta 1954), reports of this complication became frequent only after the introduction of antimetabolites in glaucoma surgery. In a review of 40 eyes with hypotony (IOP < 10 mmHg) post-trabeculectomy without antimetabolites followed for 3.4 years, Cristiansson (1967) reported persistent degenerative macular changes in four eyes, but there was no mention of the presence of chorioretinal folds or the effect of this finding on VA.

Costa et al. (1993b) reviewed the charts of 440 patients (508 eyes) who underwent trabeculectomy to detect cases of postoperative VA loss (two or more Snellen lines or a category change). In this series, six eyes (1.2%) showed loss of VA because of hypotony maculopathy. Among the six eyes, three (50%) had received postoperative applications of 5-fluouracil (5-FU) injections. Hypotony maculopathy was associated with coronary artery disease ($P = 0.0397$) and systemic hypertension ($P = 0.0118$).

Whiteside-Michel et al. (1992) evaluated the effectiveness of initial trabeculectomy with adjunctive 5-FU for uncomplicated glaucoma in 20 eyes of 20 patients younger than 40 years old. Subconjunctival injections of 5-FU (5 mg) were given 180° from the operative site within 14 days of surgery. Hypotony maculopathy was observed in one patient (5%).

Mitomycin C appears to be even more likely to produce hypotony, with the occurrence ranging from 3% to 20% in various series (Costa et al. 1993c; Kitazawa et al. 1993; Shields et al. 1993; Mégевand et al. 1995; Cheung et al. 1997; Zacharia & Schuman 1997; Perkins et al. 1998; Rasheed 1999; Lanzl et al. 2000; Martinez Garcia & Perez Garcia 2000; Mietz & Krieglstein 2001; Bindlish et al. 2002; Mietz et al. 2002; Tsai et al. 2003). In a prospective, randomized study, Rasheed (1999) compared the overall efficacy of intraoperative application of MMC in eyes with no previous ocular surgery to standard trabeculectomy without MMC. Twenty-five patients with primary glaucoma were treated with trabeculectomy without antimetabolites in one eye and trabeculectomy with MMC in the contralateral eye. After a mean follow-up of 18 months, hypotony maculopathy developed in three eyes (12%) of the MMC group, and was not observed in the group that did not receive antimetabolites.

Other studies have suggested that high concentrations of MMC or increased exposure time to MMC may be associated with the development of hypotony maculopathy (Whiteside-Michel et al. 1992; Kitazawa et al. 1993; Oppenheim & Ortiz 1993; Shields et al. 1993; Zacharia et al. 1993; Neelakantan et al. 1994; Mégevand et al. 1995). Zacharia et al. (1993) reported an incidence of 32.7% of hypotony (IOP < 5 mmHg) in 52 eyes of 48 patients undergoing trabeculectomy with MMC (0.4 mg/mL for 3.7–7 min). Twenty-two eyes requiring bilateral primary trabeculectomy were randomized by Kitazawa et al. (1993) to intraoperative MMC (0.2 mg/mL for 5 min) in one eye and to low-dose intraoperative MMC (0.02 mg/mL for 5 min) in the contralateral eye. Two cases (18.2%) of transient hypotony maculopathy were noted in the 0.2 mg/mL group exclusively. Mégévand et al. (1995) compared the results of 25 eyes that were considered to be at high risk for surgical failure.

![Fig. 8. Normal eye (A, B). Mild hypotony with thickening of the choroid, initial scleral shrinking and disc oedema (C, D). Severe hypotony maculopathy with choroidal and retinal folds, macular striae, disc oedema, dilated retinal veins and scleral shrinking (E, F).](image-url)
Table 2. Incidence of hypotony maculopathy following trabeculectomy or combined (phacoemulsification + trabeculectomy) procedures in different studies.

<table>
<thead>
<tr>
<th>Author</th>
<th>n</th>
<th>Type of surgery</th>
<th>Antimetabolites (dose)</th>
<th>Follow-up (months)</th>
<th>Incidence (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Whiteside-Michel et al. (1992)</td>
<td>20</td>
<td>TREC</td>
<td>5-FU (15-45 mg; mean 27.8 ± 8.8 mg)</td>
<td>31.1 ± 17.3 (11.5-70)</td>
<td>5%</td>
</tr>
<tr>
<td>Costa et al. (1993a)</td>
<td>39</td>
<td>TREC</td>
<td>MMC (0.4 mg/mL; 1.5-2.5 min)</td>
<td>6.7 ± 0.7</td>
<td>7.7%</td>
</tr>
<tr>
<td>Shields et al. (1993)</td>
<td>15</td>
<td>PHACOTREC</td>
<td>MMC (0.4 mg/mL; 3.5 min)</td>
<td>6.8 ± 0.9</td>
<td>7%</td>
</tr>
<tr>
<td>Kitazawa et al. (1993)</td>
<td>11</td>
<td>TREC</td>
<td>MMC (0.02 mg/mL; 5 min)</td>
<td>11.0 ± 3.4</td>
<td>0%</td>
</tr>
<tr>
<td>Mègevand et al. (2000)</td>
<td>25</td>
<td>TREC</td>
<td>MMC (0.2 mg/mL; 5 min)</td>
<td>10.7 ± 3.2</td>
<td>18.2%</td>
</tr>
<tr>
<td>Cheung et al. (1997)</td>
<td>48</td>
<td>TREC</td>
<td>MMC (0.2 mg/mL; 5 min)</td>
<td>12.0 (4-19)</td>
<td>0%</td>
</tr>
<tr>
<td>Zacharia &amp; Schuman (1997)</td>
<td>20</td>
<td>PHACOTREC</td>
<td>MMC (0.4 mg/mL; 1.0-3.5 min)</td>
<td>14.4 ± 3.1 (9.2-19.3)</td>
<td>5%</td>
</tr>
<tr>
<td>Bell et al. (1997)</td>
<td>45</td>
<td>TREC</td>
<td>5-FU (25 mg/mL; 5 min)</td>
<td>24.0 ± 6.9 (12-42)</td>
<td>4.4%</td>
</tr>
<tr>
<td>Perkins et al. (1998)</td>
<td>68</td>
<td>TREC</td>
<td>MMC (0.5 mg/mL; 0.5-5.0 min)</td>
<td>36.0 (93%)</td>
<td>5.9%</td>
</tr>
<tr>
<td>Rasheed (1999)</td>
<td>25</td>
<td>TREC</td>
<td>No antimetabolite</td>
<td>17.8 ± 1.2</td>
<td>0%</td>
</tr>
<tr>
<td>Lanzl et al. (2000)</td>
<td>10</td>
<td>TREC</td>
<td>MMC (0.4 mg/mL; 1.0-4.0 min)</td>
<td>14.9 (9-19)</td>
<td>10%</td>
</tr>
<tr>
<td>Martinez Garcia &amp; Perez Garcia (2000)</td>
<td>34</td>
<td>TREC</td>
<td>MMC (dose not available)</td>
<td>28.1</td>
<td>8.8%</td>
</tr>
<tr>
<td>Mietz &amp; Kriegstein (2001)</td>
<td>10</td>
<td>TREC</td>
<td>Suramin (200 mg/mL; 5 min)</td>
<td>18.0</td>
<td>0%</td>
</tr>
<tr>
<td></td>
<td>10</td>
<td>TREC</td>
<td>MMC (0.2 mg/mL; 3 min)</td>
<td>18.0</td>
<td>10%</td>
</tr>
<tr>
<td></td>
<td>20</td>
<td>TREC</td>
<td>No antimetabolite</td>
<td>18.0</td>
<td>0%</td>
</tr>
<tr>
<td>Mietz et al. (2002)</td>
<td>24</td>
<td>TREC</td>
<td>Postoperative MMC (0.05 mg/mL)*</td>
<td>18.2 ± 6.5</td>
<td>0%</td>
</tr>
<tr>
<td>Bindlish et al. (2002)</td>
<td>123</td>
<td>TREC</td>
<td>MMC (0.25 / 0.33 / 0.5 mg/mL; 0.5-5.0 min)</td>
<td>At least 60</td>
<td>8.9%</td>
</tr>
<tr>
<td>Tsai et al. (2003)</td>
<td>15</td>
<td>TREC</td>
<td>MMC (0.2 mg/mL; 2 min)</td>
<td>25.7 ± 21.4</td>
<td>20%</td>
</tr>
<tr>
<td></td>
<td>29</td>
<td>TREC</td>
<td>No antimetabolite</td>
<td>27.7 ± 12.2</td>
<td>0%</td>
</tr>
</tbody>
</table>

TREC, trabeculectomy; PHACOTREC, phacotrabeculectomy; MMC, mitomycin C; 5-FU, 5-fluouracil.

*Mitomycin-C (0.05 mg/mL) was applied topically to the filtering bleb on days 1, 2 and 3 after surgery (postoperative application).

and that received a single intraoperative application of MMC (0.2 mg/mL for 2 min) to a group of 48 patients – matched for age, race, type of refractory glaucoma and preoperative IOP – who received a single intraoperative application of MMC (0.2 mg/mL for 5 min). Hypotony-related maculopathy developed in one eye (2.1%) in the 5 min group.

Costa et al. (1993a) reviewed the charts of patients who underwent primary trabeculectomies (group 1, n = 39) or combined procedures (phacoemulsification + trabeculectomy – group 2, n = 15) and received intraoperative MMC (0.4 mg/mL; 1.5–3.5 min), and observed that hypotony maculopathy developed in three cases of group 1 (7.7%).

Bindlish et al. (2002) investigated the 5 year incidence of hypotony maculopathy after trabeculectomy with MMC at various concentrations. Hypotony (IOP < 6 mmHg) occurred in 42.2% of eyes after a mean follow-up of 26.1 months, whereas hypotony maculopathy occurred in 8.9% of eyes at mean follow-up of 33.7 months.

The association between hypotony maculopathy and the use of antimeabolites is not exclusively because of the antifibrotic effect, leading to reduced scar formation and a tendency for overfiltration. It has been suggested that MMC may have a direct toxic effect on the ciliary body, resulting in reduced aqueous humour production (Nuyts et al. 1994a). Additionally, the application of antifibrotic agents in filtering surgery, such as MMC or 5-FU, may induce changes in the conjunctiva. The tissue frequently becomes avascular, which prevents migration of cells via the vascular route to induce fibrosis (Shields et al. 1993; Hutchinson et al. 1994; Sihota et al. 2000). Furthermore, Sihota et al. (2000) suggested that a dysfunctional conjunctival barrier, as evidenced by the 'sweating' of the bleb and histopathologic alterations in the epithelial barrier, could be responsible for the hypotonic maculopathy in eyes undergoing trabeculectomy with MMC.

Certainly, the level of IOP alone does not determine who will develop wrinkling of the choroid and retina. Factors such as scleral thickness, scleral rigidity, structural variations in the choroid and its vessels, extraculcular muscle tone and duration of hypotony are probably important in the pathogenesis of hypotony maculopathy (Gass 1972). It is also important to notice the influence of central corneal thickness (CCT) on the diagnosis of hypotony maculopathy. Eyes with thin corneas may not develop hypotony maculopathy despite low IOPs, artificially reduced by the CCT value. On the other hand, eyes with thick corneas and IOPs of 8–9 mmHg may exhibit hypotony maculopathy despite apparently normal IOPs.

Early on, DellaPorta (1954) noticed that ocular hypotony preceded by ocular hypertension and the relatively young age of the patients involved were important prerequisites to the development of hypotony maculopathy. The medical records of 186 patients with ocular hypotony following glaucoma surgery were reviewed by Fannin et al. (2003) in a
retrospective case series to determine risk factors for hypotony maculopathy. The mean age for eyes with maculopathy was 50.7 years versus 70.7 years for those eyes without maculopathy. Maculopathy was more frequent in myopic eyes and in eyes undergoing primary surgery. Furthermore, eyes with hypotony maculopathy had a lower frequency of choroidal effusion than non-maculopathy controls.

Several authors confirmed that younger age, myopia, male gender and primary filtering surgery are risk factors for the development of hypotony maculopathy (Jampel et al. 1992; Stamper et al. 1992; Suher et al. 1997; Palmberg 1998). The first two factors support the theory of low scleral rigidity as an important element in the pathogenesis of hypotony maculopathy (Gass 1972; Stamper et al. 1992). It is possible that low scleral rigidity facilitates the inward collapse of the scleral wall during hypotony, causing the chorioretinal wrinkling. The sclera of young patients is thought to be more elastic and flexible than that of older patients, and may shrink more in hypotonic conditions. Similarly, the sclera in patients with myopia tends to be thinner, less rigid and, thus, more likely to contract in hypotony (Gass 1972).

**Differential diagnosis**

Chorioretinal folds may also develop in the absence of hypotony. The following situations may be associated with chorioretinal folds:

1. Idiopathic chorioretinal folds. Incidental finding in patients who are seen because of presbyopia and those who have normal or near-to-normal VA. These patients typically have hyperopia (1–6 dioptries or more). When the folds occur in the macular region, they are often roughly horizontal in their course or may radiate outward from the optic disc (Gass 1987).

2. Retrobulbar mass lesions. Orbital tumours as well as orbital implants, in some cases, may cause scleral oedema, choroidal congestion and chorioretinal folds (Kroll & Norton 1970; Wolter 1974; Friberg & Grove 1983). If the retrobulbar mass is removed or otherwise treated successfully, the chorioretinal folds usually disappear (Kroll & Norton 1970).

3. Scleral inflammation. Thickening and inflammation of the sclera in thyroid eye disease, inflammatory pseudotumour of the orbit and rheumatoid scleritis may cause chorioretinal folds (Coleman 1995).

4. Scleral buckle. Thickening of the sclera in the vicinity of a scleral buckle for a rhegmatogenous retinal detachment may occasionally produce chorioretinal folds (Coleman 1995).

5. Choroidal tumours. Choroidal tumours, particularly malignant melanomas and metastatic carcinomas, may produce folds in the choroid and retina. These folds are produced by mechanical displacement of the surrounding choroid by the expanding tumour (Norton 1969).

6. Choroidal neovascularization. Contraction of a choroidal neovascular membrane (CNVM) and the underlying Bruch’s membrane occurring either spontaneously or after photocoagulation may cause a radiating pattern of chorioretinal folds around the membrane (Gass 1981).

**Prevention and treatment**

The advent of antimetabolite therapy in glaucoma filtration surgery has resulted in an increased incidence of postoperative hypotony secondary to overfiltration. Modifications of the surgical technique, such as tighter scleral flap suturing and postoperative gradual increase in outflow with laser suture lysis or releasable sutures, should be used in order to avoid overfiltration and the occurrence of hypotony maculopathy (Savage et al. 1988; Melamed et al. 1990).

Hypotony maculopathy is usually treated with procedures designed to elevate the IOP. Normalization of IOP may then reverse the inward scleral bowing. However, despite successful IOP elevation, VA may not return to normal, and permanent macular chorioretinal changes may ensue (Duker & Schuman 1994).

The successful treatment of hypotony maculopathy depends on the correct identification of its cause. Once the cause is detected, treatment should be employed as soon as possible: delayed normalization of the IOP may result in permanent macular chorioretinal changes and poor vision (Costu et al. 1993c; Nuysts et al. 1994b).

Leakage of aqueous humour from a filtration bleb after glaucoma surgery may cause hypotony maculopathy. Conservative management, including the use of aqueous suppressants, pressure patching, collagen shield application or contact lens/shell/ring tamponade, may help to seal the leak (Ruderman & Allen 1985; Melamed et al. 1986; Fourman & Wiley 1989; Blok et al. 1990; Hill et al. 1990). One possible approach consists of aqueous suppressants, use of a contact lens and topical gentamicin therapy. Aqueous suppressants decrease production of aqueous humour and reduce bulk flow through the leak, thereby allowing epithelial proliferation; bandage contact lens facilitates epithelial migration; and topical aminoglycoside (e.g. gentamicin) incites mild conjunctival inflammation and stimulates wound healing (Tomlinson et al. 1987).

There have also been successful reports of bleb leaks treated with low-power argon laser application (Baum & Weiss 1993) or fibrin glue topical application (Kajiwara 1990). More recently, intrableb or subconjunctival peribleb injection of autologous blood has been described as a simple office technique for treating chronic postfiltration hypotony (Wise 1993; Smith et al. 1995) (Fig. 9).

Conjunctival compression sutures have also been used to induce adherence of conjunctiva to underlying tissues (Palmberg & Zacchei 1996; Palmberg 1996). Palmberg (1996) suggested that 9–0 nylon sutures could be employed to isolate the area of leakage, reducing the access of aqueous to the leakage site, or to limit the size of an overhanging bleb in cases of overfiltration (Fig. 10). The sutures applied to sectors of the conjunctiva in which the fibroblasts are viable may help induce sufficient healing response. However, this technique only affects the area of conjunctiva that is compressed, and, in our hands, this has been insufficient in cases where hypotony is caused by overfiltration and no leakage. When these sutures are used in association with subconjunctival injections of autologous blood, they may provide more effective management of hypotony.
maculopathy following filtration surgery with mitomycin C (Haynes & Alward 1999).

Finally, bleb excision with surgical revision, donor scleral graft patching and reconstruction of the filtering bleb with a free conjunctival autograft represent more invasive treatment modalities (Melamed et al. 1991; O’Connor et al. 1992; Buxton et al. 1994; Wilson & Kotas-Neumann 1994). These techniques are highly successful in increasing IOP by closing conjunctival leakage or decreasing filtration in overfunctioning blebs, with success rates varying from 80% to 94% (Budenz et al. 1999; Catoira et al. 2000; Burnstein et al. 2002; Bashford et al. 2004; Tannenbaum et al. 2004). However, a large proportion (up to 50%) of eyes undergoing surgery require antiglaucoma medications to control IOP, and some (up to 8%) may need new filtering procedures to reduce IOP (Budenz et al. 1999; Catoira et al. 2000; Burnstein et al. 2002; Bashford et al. 2004; Tannenbaum et al. 2004).

Burnstein et al. (2002) compared the outcomes of conjunctival advancement and non-incisional management of late-onset glaucoma filtering bleb leak in a retrospective, non-randomized trial. Fifty-one eyes of 48 patients who underwent management of late-onset glaucoma filtering bleb leaks were included. Thirty-seven eyes were included in the non-incisional treatment group and 34 eyes were included in the surgical revision group (conjunctival advancement with preservation of the preexisting bleb). The Kaplan-Meier cumulative probability of success at 24 months were 42% and 80%, respectively, for the non-incisional and surgical revision groups ($P = 0.0001$, log-rank test).

In eyes with hypotony maculopathy secondary to an overfiltering bleb, measures to incite inflammation at the bleb site are recommended to promote scarring and consequent reduction of aqueous flow. Multiple methods have been used in attempting to induce sufficient healing or fibrosis of the bleb to reverse the hypotony. It is recognized that cataract extraction performed in previously filtered eyes can cause inflammatory compromise of the filter, leading to postoperative increases in IOP (Oyakawa & Maumenee 1982; Shields 1982; Dickens & Cashwell 1996; Sibayan et al. 1997). Inflammation and scarring at the filtering site may also be stimulated by yttrium aluminium garnet (YAG) laser (Bettin et al. 1999), cryotherapy (Jampel et al. 1992; Stamper et al. 1992; Costa et al. 1993c; Nuyts et al. 1994b), diathermy (Stamper et al. 1992) and trichloroacetic acid application (Stamper et al. 1992; Nuyts et al. 1994b).

Injection of autologous blood into the bleb has been described but is not without risk. It was first employed by Wise (1993), who successfully treated four eyes with chronic hypotony following MMC trabeculectomy. Nuyts et al. (1994b) reported that 17 (77.3%) of the 22 eyes that underwent intrableb autologous blood injection showed IOPs of 6 mmHg or greater after a mean follow-up of 20.7 weeks. Leen et al. (1995) investigated the efficacy of intrableb autologous blood injection in 12 eyes with overfiltering or leaking blebs, and observed a 58.3% success rate after an average follow-up of 6.8 months. Smith et al. (1995) demonstrated a 66.6% success rate following peribleb autologous blood injection into surrounding subconjunctival tissue after a minimum follow-up of 4 months. Ellong et al.
evaluated the effectiveness of intraorbital autologous blood injections in 12 eyes with hypotony associated with overfiltration. After a mean follow-up of 12.3 months, the average IOP increased from 2.7 ± 1.2 mmHg to 8.2 ± 4.2 mmHg (P < 0.05).

Yieh et al. (2001) described the use of autologous fibrinogen concentrate (AFC) to treat seven eyes with persistent post-trabeculectomy hypotony. Under a microscope, 0.2 mL AFC and bovine thrombin was injected into the blebs. On the second day, the mean IOP of seven eyes elevated from 3.4 ± 2.1 mmHg to 12.6 ± 4.2 mmHg. Within 2 weeks, VA was noted to improve in six eyes (85.7%).

Sometimes, the above-mentioned measures used to treat hypotony secondarily to overfiltration do not reverse the problem because failure of scleral healing is the underlying cause. In these cases, resuturing the scleral flap or placing a scleral patch over the original flap is effective in reversing hypotony and restoring VA, while still maintaining some degree of filtration (Cohen et al. 1995; Schwartz et al. 1996; Halkiadakis et al. 2005; Harizman et al. 2005).

Schwartz et al. (1996) reported four eyes that underwent trabeculectomy using topical mitomycin C and developed hypotony maculopathy unresponsive to non-surgical therapies. After a minimum follow-up of 18 months, the surgical revision with scleral flap closure increased the IOP, reversing the hypotony, and resulted in improved VA to 20/25 or better in all cases.

Harizman et al. (2005) reported a modified technique of bleb revision with the use of a donor scleral patch in 15 eyes in which scleral melting did not allow effective suturing and closure of the aqueous leak. After a mean follow-up of 22.0 months, mean IOP increased from 2.9 ± 2.3 mmHg to 14.1 ± 3.3 mmHg, and mean VA improved from 20/50 to 20/30. Halkiadakis et al. (2005) employed the same technique in 14 patients. The preoperative IOP was 3.3 ± 2.6 mmHg, and the final IOP was 11.6 ± 3.4 mmHg after 10.1 ± 6.8 months. VA improved in 10 of 14 eyes. A second scleral patch graft revision was necessary in three eyes, but bleb leaks and hypotony resolved in all 14 eyes at last follow-up.

Resolution

As IOP is normalized, the choroidal folds become flattened and may completely disappear. Residual changes in the retinal pigment epithelium may remain as a result of hyperplasia and hyperpigmentation. Fluorescein angiography may depict abnormal areas of hypo- and hyperfluorescence. The choroid and the sclera recover their original thickness, and the tortuosity and engorgement of the retinal vessels disappear.

The prognosis for visual recovery apparently depends primarily on the duration of the hypotony. If retinal folds are induced by scleral shrinkage in response to low IOP, the restoration of the normal smooth architecture to the retina and Bruch’s membranes allows realignment of photoreceptors. If not treated promptly, prolonged hypotony may cause irreversible fibrosis within the retina, choroid or sclera, maintaining the choroid in a folded position (Jampel et al. 1992).

Conclusion

Hypotony maculopathy is an uncommon complication of glaucoma filtering surgery, trauma and other anterior segment surgeries. Young age (Dellaporta 1948, 1950, 1954; Jampel et al. 1992; Stamper et al. 1992; Súñer et al. 1997; Palmberg 1998), myopia (Jampel et al. 1992; Stamper et al. 1992; Súñer et al. 1997; Palmberg 1998), primary filtering surgery (Súñer et al. 1997), systemic illnesses (Costa et al. 1993b) and elevated preoperative IOP (Stamper et al. 1992; Palmberg 1998) have been found to be associated with hypotony maculopathy.

Early detection of the characteristic fundoscopic findings of hypotony and the search for its cause are fundamental because the visual deficit can be corrected through appropriate measures to restore normal IOP. The prognosis for VA recovery depends on several factors, including the duration of the hypotony. Delayed normalization of IOP may result in permanent macular chorioretinal changes and poor vision. This justifies an immediate recognition of the condition, identification of the cause and initiation of treatment.

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