

Posterior staphyloma – occurrence, classification, diagnostics and therapeutic options



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HIGHLIGHTS

Posterior staphyloma is the most destructive pathological lesion of pathologic myopia. Regular 3D-MRI and OCT are crucial for its early detection. Anti-VEGF treatment and photodynamic therapy allow for the control of selected complications, and surgical procedures show promising results, but none of these approaches is a fully satisfactory solution.

ABSTRACT

Posterior staphyloma is a key manifestation of pathological myopia, defined as a progressive bulging of the posterior eyeball wall. With increasing myopia prevalence, it remains a serious clinical problem leading to vision-threatening complications like myopic maculopathy or choroidal neovascularization. This paper reviews its pathophysiology, classifications, and diagnostic methods, highlighting ultra-widfield optical coherence tomography as a precise imaging tool. We discuss therapeutic strategies, including vascular endothelial growth factor inhibitors and posterior scleral reinforcement. Despite advances, effective strategies to inhibit progression are still needed.

Key words: posterior staphyloma, high myopia, pathological myopia

INTRODUCTION

Myopia is one of the key issues affecting the impairment of visual quality on a global scale [1]. Its prevalence has increased in recent years, particularly in Asian countries [2]. One of the factors driving this phenomenon was the COVID-19 pandemic, which contributed to children spending more time in front of screens and a decrease in outdoor activities [3]. Along with the rise in myopia, there has been an increase in the number of cases of pathologic myopia, which often occurs bilaterally in relatively young individuals [4]. Pathologic myopia is a cluster of myopic complications occurring in the posterior segment of the eye [4, 5]. One such complication is posterior staphyloma (PS) – a progressive deformation of the posterior wall of the globe, characterized by a distinct change in curvature within a localized area [6]. It is the most destructive pathological lesion of pathologic myopia, leading to more severe myopic complications [7]. The aim of this study is to summarize current knowledge on PS in the context of pathologic myopia. Particular attention has been paid to its pathophysiology, classifications, diagnostic capabilities, and therapeutic options, as well as the impact of PS on the development of myopic maculopathy and visual prognosis.

ANATOMY, PATHOPHYSIOLOGY, AND EPIDEMIOLOGY OF POSTERIOR STAPHYLOMA

The first description of PS dates back to 1801, when Antonio Scarpa documented distinct bulges in the posterior part of the eyeballs during a post-mortem examination [8, 9]. PS is a localized protrusion of the globe wall at the posterior pole, leading to deformation of the retina and choroid, as well as secondary visual disturbances [9]. From a clinical perspective, PS requires differentiation from simple posterior scleral bowing, as well as from staphylomas that have developed secondarily, for example, as a result of trauma [9]. PS most commonly develops due to high myopia and is associated with axial elongation of the eyeball, leading to scleral thinning and loss of rigidity [9]. These changes are linked to the development of myopic maculopathy; the influence of PS on the onset of myopic neovascularization and macular retinoschisis has also been described. However, PS is not exclusively associated with myopia. It also occurs in conditions such as retinitis pigmentosa, osteogenesis imperfecta, oculocutaneous albinism, and tilted disc syndrome [7, 10, 11]. The increase in patients with myopia is particularly noticeable in Asia, but the number of cases is also rising in Europe. It is projected that by the year 2050, 50% of the population will suffer from myopia, including approximately 10% from high myopia [1]. In the Chen et al. study, involving 447 eyes with high myopia, the prevalence of PS was 180 (40.27%). Among children it was 13%, in adolescents – 9%, in adults – 42.49%, and in the elderly

– 51.35% [12]. This study demonstrated that the risk of PS increases with age.

CLASSIFICATION OF POSTERIOR STAPHYLOMA

The first classification of PS was developed in 1977 by Curtin, who distinguished 10 primary and compound types in eyes with pathologic myopia [13]. It is based on the ophthalmoscopic appearance of the ocular fundus. Types I–V are considered primary staphylomas, while types VI–X are considered compound staphylomas [13]. This system was modified and simplified by Ohno-Matsui using modern imaging techniques, such as 3D-MRI [14]. This classification is based on the analysis of the entire shape of the eyeball; types VI–X were merged into a single category – type I, and the remaining types were classified according to location and distribution, which facilitates memorization [14]. The classification of PS is currently used to assess the risk and severity of maculopathy and to plan the diagnostics and treatment of patients with high myopia [8].

DEVELOPMENT OF POSTERIOR STAPHYLOMA DIAGNOSTICS

The diagnostics of PS plays a crucial role in its recognition, classification, and subsequent monitoring of complications. Over the years, imaging methods of the posterior segment of the eye have significantly evolved. New methods have provided novel information about the structure of the posterior segment of the eyeball and have gradually enabled increasingly precise examination and classification [5]. The first examination that enabled the diagnosis of PS was indirect ophthalmoscopy. In Curtin's study, involving 250 patients with myopia and PS, ophthalmoscopy allowed for the distinction of 10 types of staphyloma. The applied method enabled the determination of the location of fundus changes, their size, and the appearance of their edges [13]. Subsequently, in some cases, computed tomography (CT) was also employed. CT enabled quantitative measurements of the eyeball in various planes and comparison of its axial, vertical, and horizontal dimensions. However, a limitation remains the fact that images were analyzed primarily in 2D cross-sections, which prevents more accurate assessment. For this reason, computed tomography has not found widespread application in the routine diagnostics of PS [15, 16]. Another method used in the diagnostics of PS, which can still be considered useful, was B-scan ultrasonography. It enabled the assessment of the posterior wall of the eyeball and quantitative measurements of its deformation, which represented progress compared to previous imaging methods. In the study by Steidl et al. B-scan was used as a method allowing for objective classification of the degree of PS advancement [17]. Similarly, in the study by Park et al. B-scan

ultrasonography was also utilized as a reliable diagnostic tool [18]. Hoang et al. additionally noted that this method also enables assessment of dynamic deformations of the posterior wall of the eyeball during eye movements, which increases its diagnostic value [19]. Despite significant advantages, a limitation of this technique remains its two-dimensional nature of imaging, which hampers accurate evaluation of subtle curvatures of the posterior segment of the eyeball [5]. Magnetic resonance imaging also found application in the diagnostics of PS. However, it was initially limited to 2D imaging. Due to the two-dimensional nature of the image, the examination often did not encompass the entire width and length of the PS, which limited the possibilities of assessing its shape [20]. The application of three-dimensional magnetic resonance imaging proved to be progress, enabling analysis of the entire topography of the eye and allowing for complete visualization of changes [4, 14]. Due to high costs and limitations preventing precise assessment of the retina and choroid, magnetic resonance imaging, despite its research value, has not found application in everyday clinical practice [4, 5]. The tool that has become crucial in the diagnostics of PS is optical coherence tomography (OCT). This examination enables detailed assessment of the retina, choroid, and internal contour of the sclera [21]. With the development of technology from spectral-domain OCT to swept-source OCT, the capability of deeper penetration and better imaging of the posterior pole was achieved, which significantly broadened the clinical value of the method [22, 23]. The next step was widefield techniques. Widefield OCT allows for imaging of the entire topography of the PS. Shinohara et al. through the application of this detailed method, described 3 characteristic diagnostic features: gradual thinning of the choroid toward the PS border, internal bulging of the sclera at its edge, and its external bulging beyond the border [24]. The most recent work by Ehongo also indicates that ultra-widefield OCT currently represents the most precise method for diagnosing PS, enabling both detection of borders and monitoring of change progression [5].

COMPLICATIONS IN POSTERIOR STAPHYLOMA

PS is one of the key structural changes in eyes with high myopia. The deformation of the posterior pole of the eye itself does not necessarily cause immediate vision loss, but it significantly increases the risk of developing secondary degenerative and neovascular changes. These complications associated with the presence of PS are responsible for the gradual deterioration of visual functions in patients [7, 13]. Chorioretinal changes are among the most frequently described complications of PS and constitute a key element of the pathological myopia picture. In Curtin's study, they were found in as many as 77.5% of eyes with PS. The most characteristic changes include diffuse and focal cho-

rioretinal atrophy [13, 14, 25]. Diffuse atrophy is particularly common in type I staphylomas [25]. A relationship has also been demonstrated between the type of PS and the location of chorioretinal atrophy [26]. The picture of these changes fits into the broader definition of myopic maculopathy, classified according to the ATN system (Atrophy/Traction/Neovascularization) [7]. The presence of PS significantly increases the risk of developing all these changes and is associated with the presence of more advanced maculopathy [27]. Maculopathy associated with PS is also linked to a greater risk of visual acuity deterioration and worse prognosis [7]. Another significant complication is choroidal neovascularization (CNV), constituting the "N" component of myopic maculopathy. It can lead to sudden deterioration of visual acuity and, in the long-term course, result in the formation of a fibrous-pigmented scar referred to as Fuchs' spot [7]. Studies have shown that PS significantly increases the risk of CNV occurrence and that these changes are more frequent in patients with PS involving the macula [7]. For this reason, the presence of CNV in eyes with PS is considered one of the more important prognostic complications in pathological myopia.

VISUAL FUNCTION DISORDERS

PS leads not only to morphological changes but also to significant visual function disorders. Best-corrected visual acuity (BCVA) is significantly worse in patients with PS than in individuals with high myopia without this deformation [7, 13, 14]. Particularly unfavorable prognosis concerns PS involving the macula (types I and II), as well as complex forms, in which greater intensity of chorioretinal changes and more frequent complications are observed [7]. In the context of visual field defect examination, studies have not shown a statistically significant difference between visual field defects in patients with and without PS. However, it was noted that patients with wide macular PS experience visual field loss more frequently than patients without PS. Ohno-Matsui notes that in types VII and IX, characterized by sudden changes in scleral curvature, visual field defects progressed in 73.8% of patients during a 10-year observation period [28]. These results indicate that PS not only increases the risk of degenerative changes in the retina but also directly affects optic nerve function and visual field. Peripapillary intrachoroidal cavitation (PICC) is a rare but significant complication of PS, particularly in its peripapillary forms [29]. This change consists of the formation of cavitation spaces within the choroid in the vicinity of the optic disc, which are visible in imaging studies as hyporeflexive areas. According to the study by Ehongo et al., PICC occurred in 20.5% of eyes with PS and in 22.7% (40/176) of eyes with coexisting peripapillary staphyloma and gamma peripapillary atrophy [29]. Additionally, Ehongo et al. draw attention to the co-occurrence of PICC with the presence of myopic conus. PICC

was found in 22.7% of eyes with conus, while in the absence of conus, PICC was not observed [5, 29].

TREATMENT METHODS FOR POSTERIOR STAPHYLOMA

PS is a complication of pathological myopia that is difficult to treat. Its location in the posterior pole of the eyeball hinders treatment [5, 14]. However, taking action is crucial because PS promotes the development of complications that directly lead to progressive deterioration of vision [13]. Anti-VEGF therapy constitutes one of the treatment strategies for complications of PS when it occurs with neovascularization. In a prospective study, Doi et al. demonstrated that in patients with inferior PS and coexisting choroidal neovascularization, anti-VEGF treatment allowed for maintenance of visual acuity during a 4-year observation period [30]. This indicates that anti-VEGF therapy is effective in long-term protection of visual function in complications of PS associated with CNV, similar to other diseases with neovascularization. Data from studies on other diseases with neovascularization, such as age-related macular degeneration (AMD), confirm the long-term effectiveness of anti-VEGF therapy in maintaining or improving visual acuity [31]. However, improvement in BCVA has not been confirmed in patients with PS without neovascularization. In the study by Jun et al., the effectiveness of anti-VEGF treatment and photodynamic therapy in serous macular detachment (SRD) associated with inferior PS was evaluated. The analysis included patients without choroidal neovascularization. These results suggest that in the case of SRD in the course of PS without CNV, anti-VEGF treatment and photodynamic therapy do not provide the expected effectiveness, and the functional prognosis remains unfavorable [32]. Photodynamic therapy was one of the first methods used in treating complications of PS, particularly in cases of choroidal neovascularization. Montero et al. showed that photodynamic therapy led to CNV closure in approximately 76% of patients with high myopia [33]. Meanwhile, in the previously mentioned study, Jun et al. analyzed patients with inferior staphyloma and serous macular detachment (SRD) without CNV. It was indicated that photodynamic therapy, similar to anti-VEGF therapy in such situations, does not lead to significant and lasting improvement. In turn, the study by Ravenstijn et al. showed that photodynamic therapy did not provide significant benefits in patients with serous maculopathy induced by PS, and in most cases, only natural fluctuations or spontaneous remission were observed [34]. Vitrectomy (pars plana vitrectomy, PPV) is another approach that is mainly used in the treatment of retinal detachment in pathological myopia. Mancino et al. analyzed the results of PPV with internal limiting membrane (ILM) peeling and gas or silicone oil tamponade. In the group with gas tamponade, the anatomical success rate was 94%, and better visual

acuity improvement was achieved than in the group with silicone oil, where the primary success rate of the procedure was 54% [35]. In turn, Zhang et al. studied the effectiveness of supplementing PPV with laser photocoagulation at the staphyloma edge. In this group, the primary retinal reattachment rate was 96.7% compared to 67.7% in the standard treatment group. Additionally, the duration of silicone oil tamponade was shortened and the number of reoperations was reduced, without significant differences in visual acuity improvement [36]. Posterior scleral reinforcement (PSR) is one of the surgical techniques aimed at stabilizing the posterior pole of the eye in high myopia complicated by the presence of PS [9]. The procedure involves transplanting sclera, most often homologous or treated with chemical crosslinking, which strengthens and shapes the eyeball wall in the staphyloma region [9]. The goal is to reduce excessive scleral extensibility, shorten axial length, and improve the anatomical conditions of the macula. In the study by Zhu et al., in patients with retinal detachment associated with macular hole in high myopia, genipin-crosslinked scleral grafts were used. Improvement in BCVA was achieved, macular hole closure in 73.7% of cases, and complete retinal reattachment. A significant result was also the shortening of axial length by an average of more than 2 mm, which confirms the mechanical effect of posterior pole reinforcement [37]. The authors obtained similar results earlier in a group of patients with macular retinoschisis and macular detachment [37]. In turn, Li et al. demonstrated the beneficial effect of PSR in the pediatric population in children with unilateral high myopia. During 3 years of observation, significant shortening of axial length was observed compared to the control group. These results indicate that PSR can limit the progression of structural changes at an early stage of the disease [38]. The latest data from Ye et al. additionally confirmed that PSR in children affects not only the shortening of the eyeball axis and stabilization of visual acuity but also improves retinal microcirculation parameters assessed in angio-OCT. Increased vascular density in capillary plexuses suggests that the procedure may beneficially affect retinal metabolism, which in the future may reduce the risk of progression of degenerative changes associated with staphyloma [39]. In summary, available studies indicate that PSR may be a valuable therapeutic option in selected cases of PS and complications of high myopia. This method leads to shortening of axial length, improvement of visual acuity, and retinal stabilization, although its long-term effectiveness and impact on the progression of staphyloma itself require further observation.

EXPERIMENTAL THERAPEUTIC APPROACHES

Scleral collagen crosslinking is one of the new experimental methods aimed at strengthening the sclera and limiting

excessive eyeball elongation in high myopia. The procedure is based on the phenomenon of creating additional chemical bonds between collagen fibers, which increases its stiffness and resistance to stretching. In studies, both methods based on photosensitizers and UV-A irradiation and chemical crosslinking using genipin, glutaraldehyde, or glycer-aldehyde administered in subconjunctival injections were used [40–43]. Results obtained in animal models confirm an increase in scleral biomechanical stiffness, limitation of axial length increase, and reduction in tissue susceptibility to enzymatic degradation [40, 42, 43]. Long-term observations in rabbits showed that the scleral crosslinking effect persists for many months [41]. Despite promising results, this method has not yet been applied clinically in patients with PS. The discussed treatment methods are summarized in table 1.

CONCLUSIONS

PS is a key component of pathologic myopia leading to the development of myopic maculopathy [7–9]. Its prevalence increases with the rising incidence of myopia in the population and with the age of patients [8, 12]. Due to the diagnostic importance of PS in predicting the severity of macular lesions, regular diagnostics such as 3D-MRI and OCT are crucial for the early detection of changes [7]. Although anti-VEGF treatment and photodynamic therapy allow for the control of selected complications, and surgical scleral reinforcement procedures show promising results, none of these approaches yet represent a fully satisfactory solution. Further prospective studies with long-term follow-up are necessary to evaluate the durability of therapeutic effects and to provide a better understanding of the mechanisms underlying the formation of PS.

TABLE 1

Overview of therapeutic interventions for posterior staphyloma and related complications.

Treatment method	Primary indication	Mechanism of action
Anti-VEGF therapy [30]	myopic CNV	inhibition of vascular endothelial growth factor
Photodynamic therapy [32]	myopic CNV/serous maculopathy	laser-induced closure of neovascular vessels
Pars plana vitrectomy [35]	retinal detachment/macular hole	removal of vitreous traction and ILM peeling
Posterior scleral reinforcement [37]	axial elongation/PS progression	mechanical stabilization of the posterior pole
Scleral crosslinking [40, 41]	experimental	chemical reinforcement of collagen fibers

CNV – choroidal neovascularization; ILM – internal limiting membrane; PS – posterior staphyloma; VEGF – vascular endothelial growth factor.

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