Staphyloma: Part 1

12

Richard F. Spaide

This chapter traces the historical background of staphylomas, the possible etiology, and some problems that may be attributed to staphylomas. Chapter 13 examines the findings obtained by new imaging modalities and formulates a classification scheme.

12.1 Historical Development of Ideas

The first description of a staphyloma was made by Antonio Scarpa, a skilled anatomist who in 1801 described two eyes from a cadaver in which the posterior portion of each eye had pronounced outward bulges [1] (Fig. 12.1). Staphylomas of the anterior segment were a recognized complication of inflammation or tumors at that time, but the posterior staphyloma seemed to be different than the anterior types. In 1830, Ammon described two eyes that had expansion of the eye in the region of the fetal fissure [2]. Similar abnormalities were described in later years occurring in the context of colobomas of the iris and lens, so the fluid filled space, referred to as posterior staphyloma of Ammon, was probably a coloboma. Arlt made the connection between the posterior staphyloma of Scarpa and myopia [3, 4]. He noted myopic eyes had a conus, an area of absent choroid and retinal pigment epithelium. For the most part, a conus was only seen in eyes with myopia, although it was known that emmetropic and even hyperopic eyes could have a conus. Following the discovery by Arlt, myopia and staphyloma were thought to be synonymous, with myopia being secondary to the staphyloma. The presence of a staphyloma was inferred to be present in myopic eyes by seeing the presence of a conus. (This was at a time when there was not widespread use of binocular ophthalmoscopy.) Arlt reasoned the conus was the result of atrophy of the choroid but seemed to have a harder time trying to determine the retinal abnormalities that accompanied the conus. Arlt knew that eyes with a conus had a larger blind spot and therefore must have had a localized absence of light sensitive cells. Other competing ideas were a conus was due to inflammation or was congenital.

Through extensive analysis of myopic eyes, Tscherning in 1883 determined that many eyes with myopia did not have a staphyloma [5]. In 1898, Schnabel reviewed the published cases and added additional new information [6]. He found staphylomas only in eyes with greater than 8 diopters of myopic refractive error. These eyes nearly always had a conus, although a conus could frequently be found in eyes with less than 8 diopters of myopic refractive error. Schnabel thought the conus was present from early in life but, with ocular expansion due growth and myopia, became increasingly evident. The divergence in the early ideas about the conus carried through to ideas of how staphyloma formed. One was atrophy of the choroid along with "the strain of near work" produced a staphyloma in an otherwise normal eye [7]. A second line of reasoning was the sclera was abnormal due to defective development and later stretched to cause a staphyloma. Another hypothesis was localized inflammation caused anterior staphyloma as well. However, Knowles discredited this idea by noting a lack of objective signs of inflammation in most of these eyes [8]. The definition of a staphyloma was codified over time. An ectasia was an outpouching of the wall of the eye without uveal tissue. A staphyloma was an outpouching of an eye with associated uveal tissue. These definitions continue to have weaknesses in that in advanced stages of myopic degeneration, the choroid within the region of the staphyloma can appear to completely regress leaving what is called chorioretinal atrophy. Despite the absence of uveal tissue, the area is still called a staphyloma.

12.2 The Curtin Classification

Curtin greatly expanded the clinical understanding of staphyloma formation in myopes [9, 10]. Curtin divided the ophthalmoscopic appearance of a group of 250 patients with

R.F. Spaide, MD
Vitreous, Retina, Macula Consultants of New York,
460 Park Ave, 5th Floor, New York,
NY 10021, USA
e-mail: rickspaide@gmail.com

R.F. Spaide et al. (eds.), *Pathologic Myopia*,
DOI 10.1007/978-1-4614-8338-0_12, © Springer Science+Business Media New York 2014

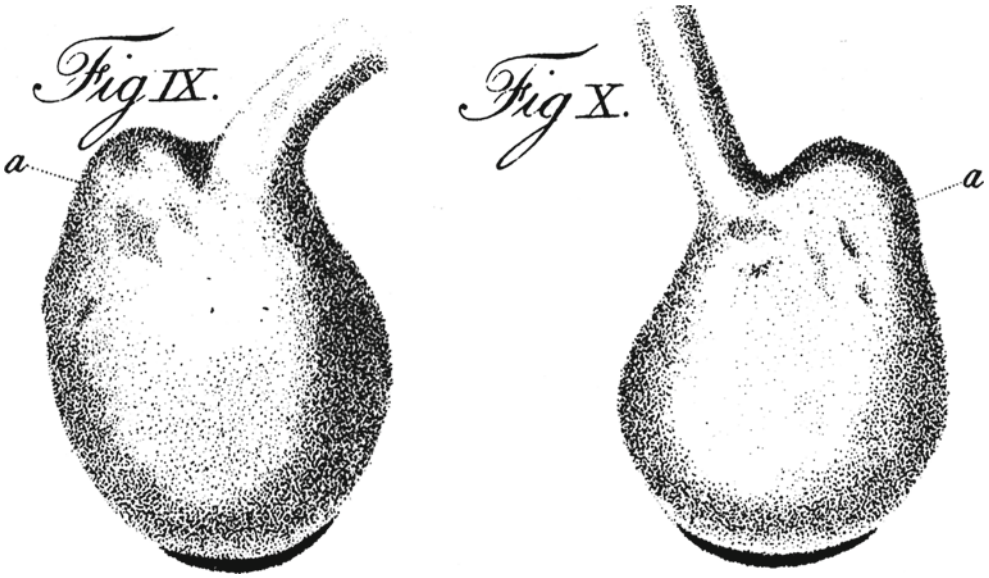

Fig. 12.1 From Scarpa [1]: "…I have twice happened to meet with the staphyloma of the sclerotic coat in its posterior hemisphere, in the dead subject, where I do not know that it has been seen or described by any other. The first time was in an eye taken from the body of a woman 40 years old, for another purpose. [In reference to Figure IX.] This eye was of an oval figure, and upon the whole, larger than the found one of the opposite eye. On the posterior hemisphere if this eye, and on the external side of the entrance of the optic nerve, or on the part corresponding to the temple of that side, the sclerotic was elevated in the form of an oblong tumor of the size of a small nut. [In reference to Figure IX, a.] When the posterior hemisphere of the eye was immersed in spirit of wine, with a few drops of nitrous acid added to it, in order to give the retina consistence and opacity, I could perceive distinctly, that there was a deficiency of the nervous expansion of the retina within the cavity of the staphyloma; that the choroid coat was very thin and discolored at this part, and wanted its usual vascular plexus; and that the sclerotic, particularly at the apex of the staphyloma, was rendered so thin as scarcely to equal the thickness of writing paper. I knew that the woman from whom the eye had been taken, had lost the faculty of seeing on the side some years before, during an obstinate ophthalmia, attended with a most acute and almost habitual pain in the head. [Figure X in the same text was from an eye contributed to Scarpa]"

staphylomas into ten different patterns (Fig. 12.2). The first five were posterior outpouchings, which Curtin called simple staphylomas, and involved the macula and optic nerve region or were centered on the macula, directly over the optic disc, nasal to the optic disc, or inferior to the optic disc [10]. Curtin showed the inferior staphyloma to be associated with a tilted optic disc, a condition that will be described in greater detail. However, other staphyloma types may also be associated with a tilted disc appearance. The remaining five were termed compound staphylomas, and these appeared to be composites of more than one type of staphyloma or were an elaboration from simple staphylomas. For example, a Type VI staphyloma is a combination of a Type I, a staphyloma over the posterior pole along with a Type II staphyloma, which is the one centered on the macula. This creates a two-tiered depression. Type IX staphylomas are two adjacent staphylomas, one principally involving the disc and the second, the macula. Many of these types of staphyloma can be associated with a tilted disc, although this was not mentioned. The Curtin classification is not exhaustive of all types of staphyloma. Many of the eyes imaged by Moriyama and associates with high-resolution magnetic resonance imaging (MRI) and 3D rendering had staphyloma configurations not anticipated by Curtin [11]. The classification scheme a landmark though, because it implicitly considered a staphyloma to be a particular anatomic configuration in the context of the overall structure of the eye. Whereas Alrt considered the staphyloma to be myopia, and vice versa, Curtin's classification highlighted the specific structural abnormalities that sometimes could be seen in eyes with increasing amounts of myopia.

The distinction of a staphyloma as being a separate sequela of myopia as opposed to being an integral part of myopia is not universally accepted. It is still common for authors to refer to abnormalities in the posterior pole of myopic eyes, even the ones that do not involve outpouching, as being staphylomas [12]. The curvature of the posterior pole of the eye in high myopes has been called a posterior staphyloma without evidence being shown that there was an alteration in the generalized curvature of the eye or any semblance of an outpouching [13]. Part of the confusion is related to how the axial length of the eye may increase as a consequence of a staphyloma. In that case, the incremental exaggeration in the refractive error of the eye is due to the increase in axial length caused by the staphyloma. Much of the confusion concerning staphylomas is due to the lack of an unambiguous and detailed definition for staphyloma beyond the rather nebulous concept of a "posterior outpouching" of the eye.

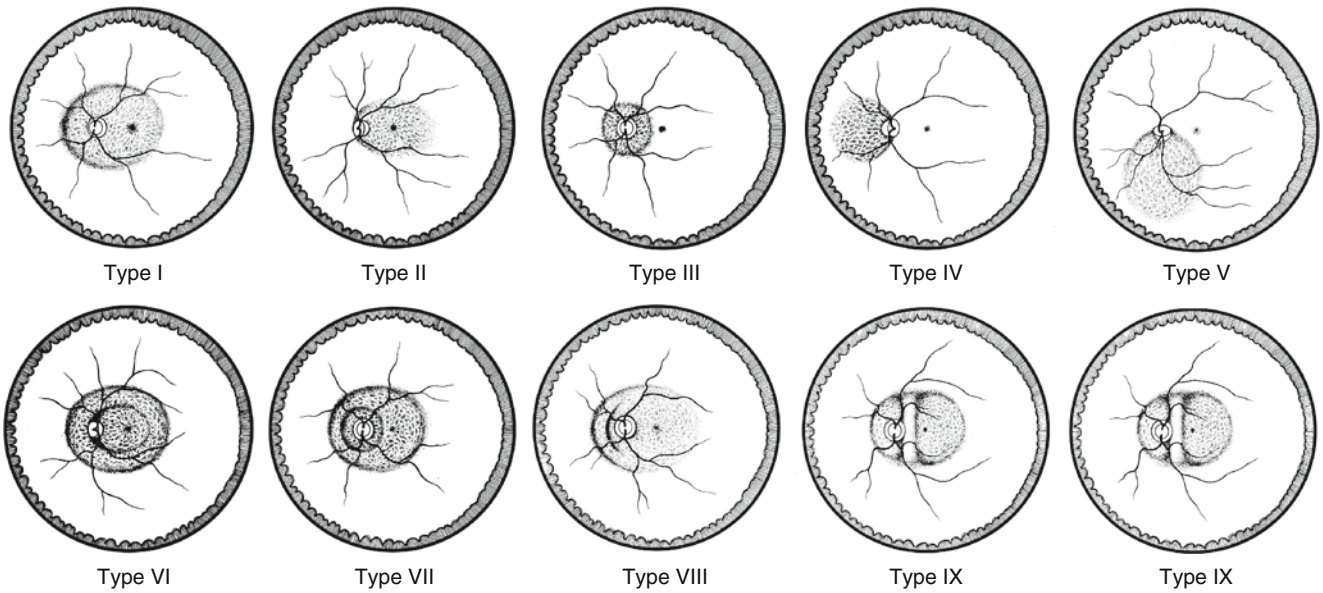

Fig. 12.2 Curtin classified the staphylomas he found in myopes into ten categories. The first five were simpler configurations, while the last five were either more intricate in their configuration [10]

Curtin used ophthalmoscopy and fundus drawings to classify the staphyloma types, while other studies employed color fundus photographs or a combination of ultrasonography and indirect ophthalmoscopy. Curtin found the most common type of staphyloma involved the posterior pole in general, causing an outpouching of the region encompassing the nerve and macula. This staphyloma type accounted for approximately one-half of all staphylomas seen in a series of 250 affected patients [10]. Curtin's Type II staphyloma, which is centered on the macula, was the second most common type seen in patients aged 3–19 years but became less common in older individuals. More complex forms of staphyloma seen in older patients were Type VIII through Type X. Type VIII is a combination of a Type 1 and a Type II. It is possible that the peculiar anatomy of the scleral collagen fibers affects the configuration observed. In the posterior portion of the eye, some of the scleral fibers form annular rings around the scleral opening for the nerve. It is possible that selective alteration of the scleral fibers could allow some of the annular fibers to remain relatively unaffected, thus producing a septum in the middle of what would have been a larger Type II staphyloma to produce a Type IX staphyloma. A Type X staphyloma is a modification of a Type II but with a shallow temporal border and an incomplete border around the optic nerve.

The staphylomatous process results in a localized thinning of structures in the posterior pole and potentially increases the likelihood of ocular abnormalities. In reality, the abnormalities seen in high myopes are highly correlated with axial length but may be exaggerated in posterior staphylomas. There are some types of staphyloma that confer special risk for ocular abnormalities, and these will be discussed later.

12.3 Prevalence of Staphyloma

The prevalence of staphylomas in highly myopic eyes could be expected to vary based on the composition of the group being evaluated in regard to axial length and method of patient recruitment. Patients being randomly selected from a pool of high myopes may have a different proportion of staphylomas than a group attending an eye clinic. The method of detecting staphylomas highly influences what is considered a staphyloma. Curtin and Karlin used ophthalmoscopy to examine eyes but did not state what they considered a staphyloma to be [10]. Hsiang et al. used ultrasonography to evaluate eyes; they measured the depth of the eye posterior to the optic nerve [14]. This method axiomatically considers eyes with staphylomas only involving the optic nerve to have no staphyloma, since in these eyes, the optic nerve is the most posterior part of the eye. In an eye with a spherical shape and the fovea in the optic axis, the fovea by default would be posterior to the optic nerve; therefore, a normal eye would be considered to have a staphyloma, albeit small, using this methodology.

Estimates of staphyloma prevalence therefore are more of a guide than an absolute. Curtin and Karlin found the prevalence increased dramatically with elongated axial length. Curtin and Karlin determined the prevalence of posterior staphyloma increased from 1.4 % in eyes 26.5–27.4 mm to 71.4 % in eyes having an axial length from 33.5 to 36.6 mm. The mix of the type of staphyloma varied with age. Younger eyes had a predominance of simple forms, with staphylomas involving either the entire posterior pole or just the macular region accounting for nearly all staphylomas seen. In older

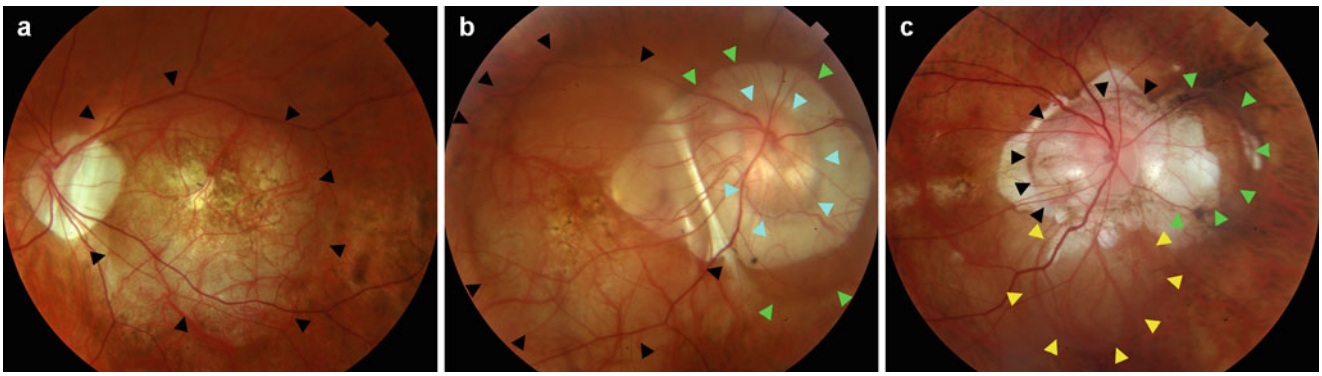

Fig. 12.3 Three different configurations of staphyloma are shown. (**a**) A staphyloma involving the macula region with its nasal border abutting the optic nerve. This would probably be close to what Curtin called a Type II staphyloma, but note that the optic nerve is tilted in this case. The posterior pole shows generic effects of the staphyloma; there is thinning of the choroid that is more pronounced in the staphyloma with overlying pigmentary changes. (**b**) The patient appears to have two staphylomas adjacent to each other (*black and green arrowheads*) and therefore would be most similar to a Type IX staphyloma as described by Curtin. However, close inspection reveals another staphyloma (*light blue arrowheads*) contained in the staphyloma around the nerve (*green arrowheads*). So this eye would have a combination of a Type III and a Type IX staphyloma. (**c**) This patient has three adjacent staphylomas (*black, green, and yellow arrowheads*) and therefore does not resemble any of the staphyloma types described by Curtin

eyes, the proportion of more complex staphylomas increased. Hsiang et al. using ultrasonography determined that staphylomas were present in 90 % of a group of 209 eyes with high myopia. They too found the complexity of staphylomas increased with advancing patient age [14].

12.4 Proposed Nomenclature

A highly myopic eye may have shape distortion where the curvature of the eye deviates from the flattened sphere seen in emmetropes without having a staphyloma. The barrel-type shape as described by Moriyama and coauthors based on 3D MRI is an example of a shape distortion that does not necessarily have staphyloma formation [11]. A staphyloma can be given a formal definition of an outpouching of the wall of the eye that has a radius of less than the surrounding curvature of the wall of the eye. A simple staphyloma is a region that has only 1 radius of curvature. A complex staphyloma is an expansion that has two distinct radii of curvature with a total or partial overlap in the curves. A compound staphyloma is present when there are two or more separate outpouchings that are not concentric. Thus, what Curtin called a Type VII coloboma, which is a staphyloma around the optic nerve embedded in a staphyloma involving the posterior pole, would be considered a complex staphyloma. The Type IX, which is two staphylomas side by side, would be a compound staphyloma. As shown in Fig. 12.3, it is relatively easy to find staphylomas that do not fit into Curtin's classification system. As such a description of the staphylomas using this proposed nomenclature would prove to be more accurate. Many of the eyes shown by Moriyama et al. [11] appear to have compound staphylomas in arrangements not anticipated by the Curtin classification. While the nomenclature above provides an accurate way to describe changes, a classification scheme also needs to have simplicity and clinical applicability to be useful.

12.5 Etiology

The enigma of the staphyloma is wrapped in the mystery of myopia. The ocular shape experiments in animal models of emmetropization show the eye is capable of local reaction to defocus [15–24]. An error signal appears to be created, and the eye is capable of responding by regional growth changes to minimize the error signal [17, 24]. The defocus induced can arbitrarily be selected by using eyeglass lenses to induce a compensatory hyperopia or myopia. This implies the amount and direction of the error signal is correctly interpreted in the response by the eye [24]. Removing the lens inducing a refractive shift from the eye of a chick results in a recovery of the eye with reduction or elimination of the induced refractive error. Obscuring vision in the growing eye sets off a related series of events that leads to myopia formation. In this situation, the emmetropization process seems to be clamped into producing one of the two potential extremes, myopia.

In animal models, there appear to be two main modalities at play in ocular adaptation to defocus. In bird, there can be a large change in the thickness of the choroid. The increase in thickness is accomplished with the aid of the lymphatic system found in bird eyes [25]. Myopization is associated with decreased choroidal blood flow and potential for choriocapillaris changes even in the short term [26, 27]. Humans, of course, have no lymphatic system, but defocus can induce a small change in the thickness of the choroid as determined by optical low-coherence reflectometry [28]. Longer-term alterations in emmetropization are related to increase in axial

length caused by expansion of the eye. The sclera undergoes remodeling, thinning, and develops increased elasticity. In animal models with optically imposed defocus in one hemifield produced alteration in the size of that hemifield in particular [29, 30]. This suggests there are mechanisms that afford local and selective control of eye growth and refractive development. Subsequently, the role of the entire retina in producing axial lengthening was studied. Interestingly, foveal ablation in the monkey followed by form deprivation still causes myopia with increased axial length [31]. Relative peripheral hyperopic defocus causes axial length changes, leading to the suggestion that peripheral optical approaches may be a potential strategy in the prevention and treatment of myopia [18, 32].

The major component of the sclera is Type I collagen. These form fibrils that provide structural strength of the eye. There is a gradient in scleral fibril size from the inner to outer sclera, and this gradient follows the development of the sclera, which progresses from the inner side to the outer surface. In high myopes, there is a decrease in the dry weight of collagen as compared with emmetropes, and the fibril diameter is decreased as well. The sclera of high myopes does not show the typical gradient of fibril diameters, as all of the fibrils are small in diameter. This change is most evident in the posterior portion of the eye, even in the absence of a staphyloma [33, 34]. There are decreased glycosaminoglycans as compared with emmetropic eyes as well. In animal models of myopia, the changes in the sclera, manifested by scleral thinning, are the results of a loss of tissue and stretching. The thinning of the sclera, along with some increase in elasticity [35], results in increased stretching (strain) for any given stretch (stress). By Laplace's law, the stress on the sclera can be estimated by $\delta = PR/2T$, where δ is the stress, P the intraocular pressure, R the radius, and T the wall thickness. In high myopes, the wall stress increases with expansion of the eye and decreased wall thickness.

In most adults, the refractive error and axial length remain stable over time. In high myopes, a subset of the population, both axial length and refractive error, can increase over time, even in adults [36, 37]. While there may be some weak genetic influence for the development of high myopia, for the most part, this disorder appears to be inducible in susceptible populations. This is evidenced by epidemiologic studies showing near work and lack of outdoor activities being the strongest risk factors, by the appearance of myopia in agrarian and hunting populations that began schooling, and by animal experimentation [38–48]. This implies there is a change in highly myopic eyes that confers increase risk for progressive changes in the architecture of the sclera, not only during phases of rapid growth, such as in late childhood, but much later in life. Given that emmetropization appears to involve local control, it is possible that regional defocus of images could cause regional alterations in scleral growth and mechanical properties. Outdoor activities involve distances that are generally greater than 1 m from the observer. That means a range of refractive differences that at most can be 1 diopter. In the bright illumination of sunlight, small pupil sizes lead to an increased depth of field, reducing the defocus of images even more. On the other hand, the range of distances from the eye varies substantially indoors; some objects may be centimeters away, while others are meters away. These distances range across a large breadth of diopters. Illumination levels are lower indoors, and the spectral composition of light may be more myopigenic [49]. In this case, it is easily possible that regions of the eye can be in focus, while other areas are defocused. Indeed one of the strongest factors related to global expansion is the amount of peripheral, not central, refractive error. Results of animal models strongly implicate image defocus in the response of regional adaptive mechanisms in the eye. The influence that prolonged defocus of images has on the development of staphylomas in humans is not known. However, in animals defocus is a commonly used method to cause myopia. If this same pathogenic mechanism exists in humans, localized defocus could cause regional expansion of the eye. It is possible that various regions of the eye wall have varying susceptibility to ocular expansion in the adult human eye leading to nonuniform expansion.

A second but related possibility is that choroidal abnormalities may play a role in the development of nonuniform expansion of the eye. This intriguing possibility is suggested by the observation of rapid and profound ocular expansion with the development of high myopia in an adult following resolution of Vogt-Koyanagi-Harada disease in which there was marked thinning of the choroid seen. The scleral composition and shape has been theorized to be controlled in part by the choroid. In the development of high myopia, the choroid becomes quite thin and in some regions disappears altogether. Any potential mechanisms involved in maintenance of ocular shape and size could potentially be altered.

12.6 Special Problems in High Myopes That Can Be Attributed to Staphylomas

The stretching of the ocular layers in a staphyloma is locally exaggerated within a staphyloma. The adverse effects of any stretching may therefore be worsened within the confines of the staphyloma. The change in contour of the eye wall may also cause potential abnormalities if the inflection point is somewhere near the macula. This is exactly what happens in a Type V staphyloma, also known as an inferior staphyloma or as tilted optic disc syndrome.

Type V staphylomas are unusual in that the center of the macula can be bisected by the change in curvature of the upper edge of the staphyloma. The superior half is less

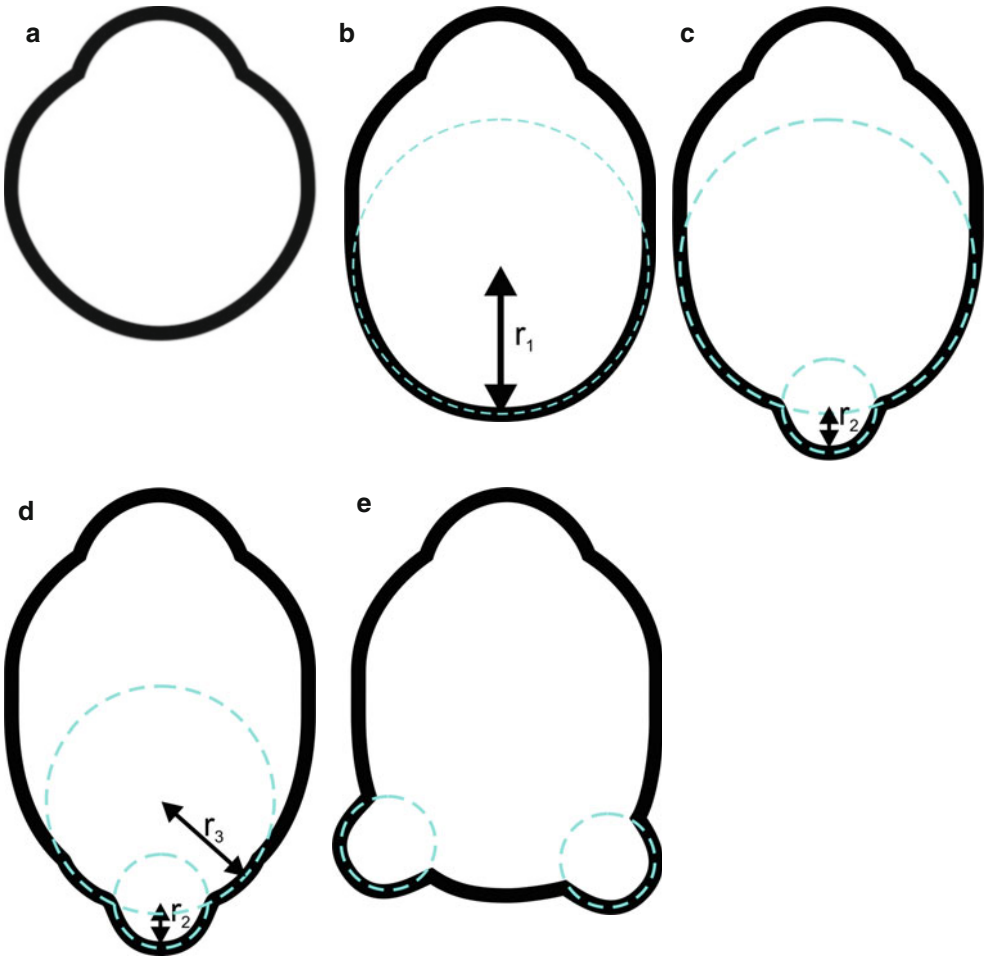

Fig. 12.4 Proposed nomenclature for staphylomas. (**a**) Normal eye shape. (**b**) Axial expansion occurring in the equatorial region that does not induce any altered curvature in the posterior aspect of the eye. This eye would have axial myopia but no staphyloma. (**c**) A second curvature occurs in the posterior portion of the eye, and this second curvature has a smaller radius (r_2) than the surrounding eye wall (r_1). This secondary curve is a staphyloma and because there is only one additional curvature is considered to be a simple staphyloma. (**d**) Some eyes have a staphyloma within a staphyloma or a complex staphyloma. Note the two secondary radii, r_2 and r_3. (**e**) Other eyes may have more than one nonassociated secondary curvatures. This is an example of a compound staphyloma

distended than the inferior portion. The nerve is usually at the border of the staphyloma and has a tilted appearance. This has led to two different names for this structural configuration: inferior staphyloma or tilted optic disc syndrome, which was the first term used to describe the disorder [50– 62]. There are numerous abnormalities described in association with this particular staphyloma. The inflection of the curve of the eye wall to the staphyloma has been associated with a band of decreased pigmentation extending in an arcuate manner along the upper edge of the staphyloma [60]. Some eyes show more complicated pigmentary patterns. The underlying choroid is thin and the sclera under that is thickened as compared with nearby areas. It is not uncommon for these eyes to have subretinal fluid [53]. There are numerous potential theories as to the origin of the fluid: the atrophy of the retinal pigment epithelium may decrease its pumping function or the scleral thickness may impede the uveal-scleral outflow there [62]. Some patients seem to have leaks seen during fluorescein angiography consistent with central serous chorioretinopathy, which is distinctly uncommon in high myopes. Some authors have proposed the subretinal fluid caused the observed retinal pigment epithelial changes [62]. Tilted disc syndrome has been associated with an uneven distribution of drusen in which the superior portion had more drusen than the inferior more myopic aspect [57]. Polypoidal choroidal vasculopathy has been described in several eyes with tilted disc syndrome [55, 56]. Typical choroidal neovascularization, including cases with many foci, has been reported in these eyes [52]. It is possible that the region of the change of curvature has numerous microbreaks of Bruch's membrane. Eyes with tilted disc syndrome commonly have superior visual field defects, which can be ameliorated by changing the refraction used while measuring the visual field [58]. This appears to be related to the disparity in axial length, although there may be neurogenic or retinal contributions to the field defect in some patients.

The exaggerated effects of thinning and curvature of the eye can induce local effects in the staphyloma (see Fig. 12.3a). The more subtle effects of decreasing choroidal thickness on visual acuity may come into play within a staphyloma if the choroidal thickness is severely compromised. The outpouching of the eye also causes the surface area occupied within that region to be greater than what it would have been for structures lining the wall of the eye. For example, the retina is stretched over a larger area increasing the stress on the retina, which is elastic to a certain extent. Because of the curvature of the eye and of the

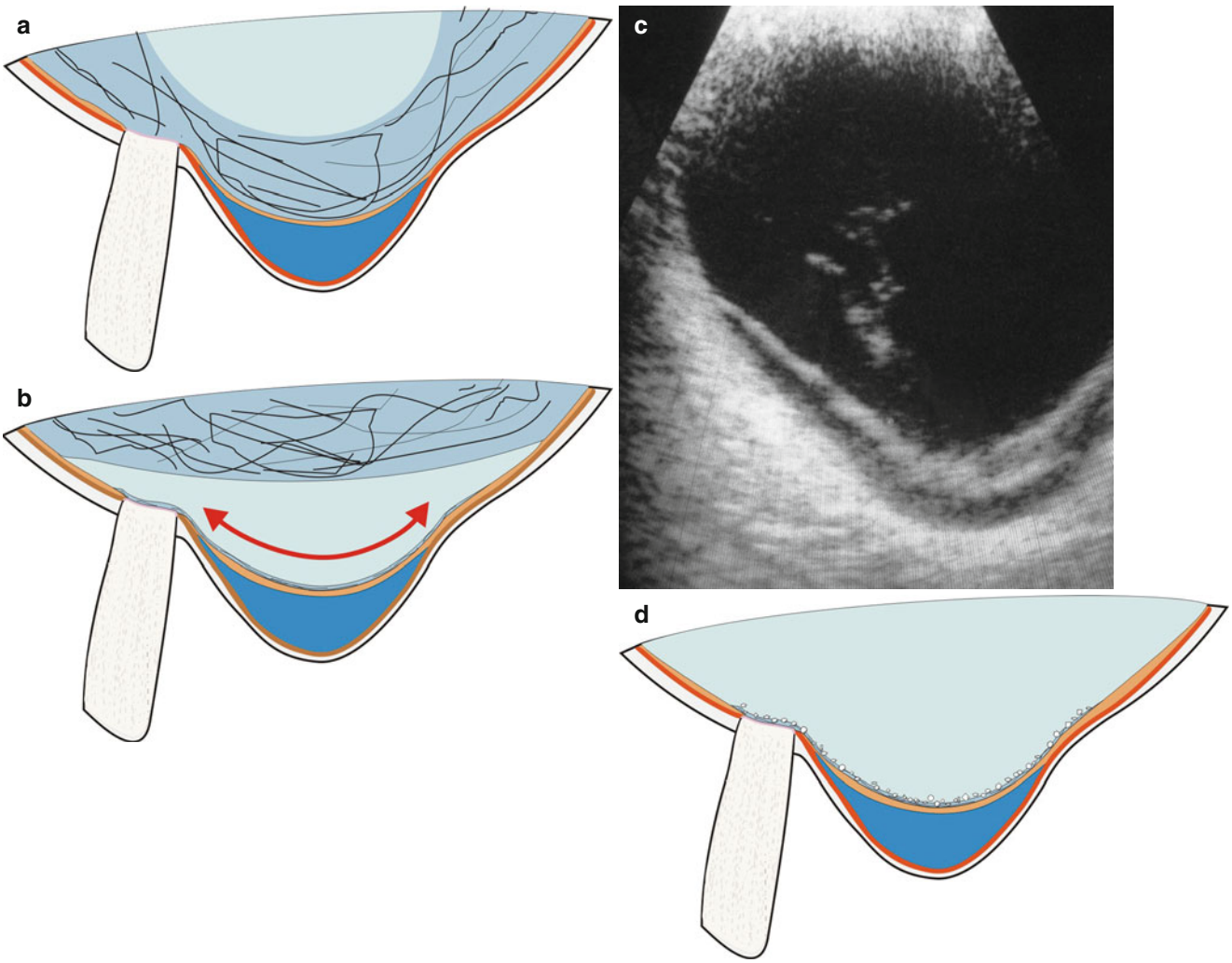

Fig. 12.5 The posterior extent of the sclera and choroid bulge backward in a staphyloma. The forces leading to attachment include the net vector of flow that goes from the vitreous to the choroid and the pumping effect by the retinal pigment epithelium. The forces that lead to detachment include traction by the attached vitreous (**a**) or even adherent plaques of vitreous (**b**) after posterior vitreous detachment. The natural elasticity of the retina also is an important factor. The retina is usually taut across the staphyloma with the apparent forces in the plane of the retina (*double arrow* in **b**). This force can be resolved into two vectors, including one that is perpendicular to the retina leading to the center of the eye. (**c**) An illustrative contact b-scan ultrasound is shown of an eye with a localized detachment in a staphyloma. (**d**) Removal of vitreous traction can include using triamcinolone (shown as white crystals on the surface of the vitreous) and possible removal of the internal limiting membrane as well

staphyloma, the forces generated by the retina can be resolved into vectors acting in the plane of the retina and perpendicularly inward from the retina (Fig. 12.4). This second vector has the propensity to lift the retina from the back wall of the eye. Any local traction on the retina through epiretinal membrane or remnants of attached vitreous would increase this vector. Counteracting this vector would be the normal forces maintaining attachment. These include the pumping by the RPE and the normal vector of flow from the vitreous to the choroid [63]. The vector of forces directed perpendicularly inward can begin to exceed the tensile strength of the retina or the force of retinal attachment. This can lead to retinoschisis or retinal detachment, respectively. Surgical repair of the induced problems can include vitrectomy with removal of the plaque of vitreous alone or with removal of the internal limiting membrane (Figs. 12.5 and 12.6) [63].

Staphyloma formation may place the fovea at an angle to the optical axis of the eye, with the outer segments discs not being perpendicular to the incoming light rays. This has the potential of inducing the Stiles-Crawford effect; the first type is a reduction in sensitivity and the second type an alteration in the perceived wavelength of the light. Asymmetrical Stiles-Crawford patterns have been associated with myopic abnormalities of eye shape [64]. High myopes have altered color vision and dark adaptation, although there are numerous possible explanations for these findings [65].

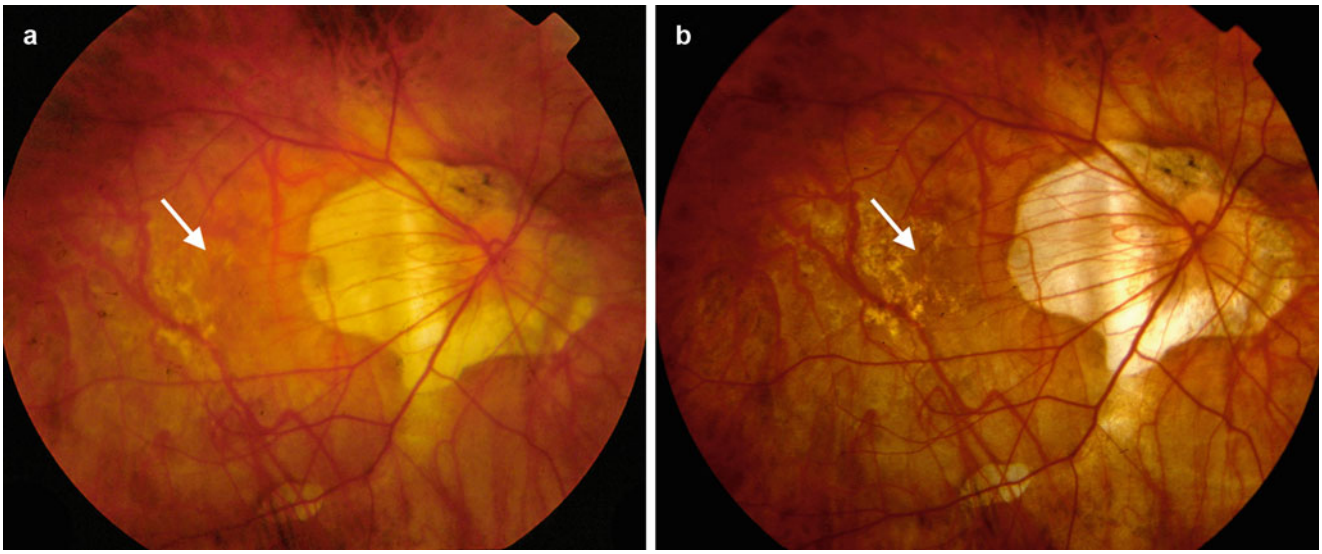

Fig. 12.6 (**a**) This patient has a shallow detachment of the retina over a staphyloma. Note the lack of visibility of the underlying choroidal details (*arrow*). (**b**) After vitrectomy and flattening of the retina, the visual acuity improved. Note the increased visibility of the underlying details (*arrow*)

12.7 A Distinct Alteration in Scleral Contour: Dome-Shaped Macula

Gaucher and colleagues described a new entity in eyes with high myopia [12]. The central portion of the macula appeared to bow inward, unlike typical staphylomas in high myopia in which staphylomas bow outward. They called the disorder dome-shaped macula in eyes with myopic staphyloma. The authors proposed that there was localized thickening of the choroid as a possible explanation. They did not demonstrate any outpouching of the eye. Later in a letter to the editor, two additional possibilities were suggested: the first was the dome shape was due to vitreous traction, and the second was there really was collapse of the posterior portion of the eye such that the sclera bowed inward [66]. The hypothesis that there was vitreous traction was not borne out by any of the OCTs, and the eyes with dome-shaped macula have normal intraocular pressure, making eye collapse highly implausible. Imamura and associates examined a group of 15 patients (23 eyes) with dome-shaped macula with EDI OCT [67]. The mean age of the patients was 59.3 years, and the mean refractive error was −13.6 diopters. The mean subfoveal scleral thickness in 23 eyes with dome-shaped macula was 570 μm, and that in 25 eyes of myopic patients with staphyloma but without dome-shaped macula was 281 μm ($P < 0.001$) even though both groups had similar refractive error. The scleral thickness 3,000 μm temporal to the fovea was not different in the eyes with dome-shaped macula, 337 μm as compared to the eyes without dome-shaped macula, 320 μm. Dome-shaped macula appears to be the result of regional thickness differences of the sclera in highly myopic eyes, and it does not correspond to any of the known types of staphyloma described (Fig. 12.7). Some patients with dome-shaped macula may have focal collections of subretinal fluid, which may occur because of the obstruction of outflow of choroidal fluid by the local scleral thickness variation

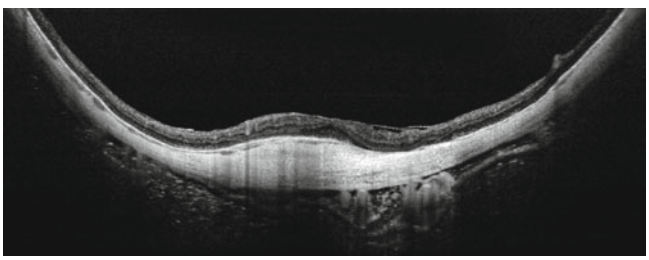

Fig. 12.7 This is a 12 mm scan obtained with a swept source optical coherence tomography instrument of a patient with a dome-shaped macula. Note that lack of any "outpouching" of the eye, meaning there was no demonstrated staphyloma. However, the posterior sclera was thicker than the more peripheral portions, although both the subfoveal and peripheral wall were thinner than an emmetrope

References

1. Scarpa A. Chapter 17. Dello Stafiloma. Practical observations on the principal diseases of the eyes. Pravia: Presso Baldassare Comino; 1801. p. 215–28.
2. Lawrence W. Section III. Staphyloma scleroticae. In: A treatise of the diseases of the eye. 3rd ed. London: Henry G. Bohn; 1844. p. 337–9.
3. Arlt F. Die Krankenheiten des Auges fur praktische Artze. Prague: F.A. Credner; 1859.
4. Arlt F. Über die Ursachen und die Entstehung der Kurzsichtigkeit. Vienna: Wilhelm Braumueller; 1876.

5. Tscherning M. Studien über die Aetiologie der Myopie. Graefes Archive for Clinical and Experimental Ophthalmology. 1883;29: 201–72.
6. Schnabel I. The anatomy of staphyloma posticum, and the relationship of the condition to myopia. In: Norris WF, Oliver CA, editors. System of diseases of the eye, Local diseases, glaucoma, wounds and injuries, operations, vol. 3. Philadelphia: J.B. Lippincott Co; 1898. p. 395–411.
7. Souter WN. Posterior staphyloma in the refraction and motility of the eye. For students and practitioners. Philadelphia: Lea Brothers & Co; 1903. p. 249–55.
8. Knowles RH. An encyclopedia-dictionary and reference handbook of the ophthalmic sciences. New York: The Jewelers Circular Publishing Company; 1903.
9. Curtin BJ, Karlin DB. Axial length measurements and fundus changes of the myopic eye. Part 1. The posterior fundus. Trans Am Opthalmal Soc. 1970;68:312–34.
10. Curtin BJ. The posterior staphyloma of pathologic myopia. Trans Am Ophthalmol Soc. 1977;75:67–86.
11. Moriyama M, Ohno-Matsui K, Modegi T, et al. Quantitative analyses of high-resolution 3D MR images of highly myopic eyes to determine their shapes. Invest Ophthalmol Vis Sci. 2012;53(8): 4510–8.
12. Gaucher D, Erginay A, Lecleire-Collet A, et al. Dome-shaped macula in eyes with myopic posterior staphyloma. Am J Ophthalmol. 2008;145:909–14.
13. Ikuno Y, Tano Y. Retinal and choroidal biometry in highly myopic eyes with spectral-domain optical coherence tomography. Invest Ophthalmol Vis Sci. 2009;50(8):3876–80.
14. Hsiang HW, Ohno-Matsui K, Shimada N, Hayashi K, Moriyama M, Yoshida T, Tokoro T, Mochizuki M. Clinical characteristics of posterior staphyloma in eyes with pathologic myopia. Am J Ophthalmol. 2008;146(1):102–10.
15. Young FA. The effect of nearwork illumination level on monkey refraction. Am J Optom Arch Am Acad Optom. 1962;39:60–7.
16. Shen W, Vijayan M, Sivak JG. Inducing form-deprivation myopia in fish. Invest Ophthalmol Vis Sci. 2005;46(5):1797–803.
17. Wallman J, Gottlieb MD, Rajaram V, Fugate-Wentzek LA. Local retinal regions control local eye growth and myopia. Science. 1987;237(4810):73–7.
18. Smith 3rd EL, Hung LF, Huang J. Relative peripheral hyperopic defocus alters central refractive development in infant monkeys. Vision Res. 2009;49(19):2386–92.
19. Schaeffel F, Glasser A, Howland HC. Accommodation, refractive error, and eye growth in chickens. Vision Res. 1988;28:639–57.
20. Smith 3rd EL, Hung LF. The role of optical defocus in regulating refractive development in infant monkeys. Vision Res. 1999;39: 1415–35.
21. Graham B, Judge SJ. The effects of spectacle wear in infancy on eye growth and refractive error in the marmoset (Callithrix jacchus). Vision Res. 1999;39:189–206.
22. Norton TT, Siegwart JT, Amedo AO. Effectiveness of hyperopic defocus, minimal defocus, or myopic defocus in competition with a myopiagenic stimulus in tree shrew eyes. Invest Ophthalmol Vis Sci. 2006;47:4687–99.
23. Shen W, Sivak JG. Eyes of a lower vertebrate are susceptible to the visual environment. Invest Ophthalmol Vis Sci. 2007;48:4829–37.
24. Zhu X, Park TW, Winawer J, Wallman J. In a matter of minutes, the eye can know which way to grow. Invest Ophthalmol Vis Sci. 2005;46(7):2238–41.
25. Nickla DL, Wallman J. The multifunctional choroid. Prog Retin Eye Res. 2010;29(2):144–68.
26. Fitzgerald ME, Wildsoet CF, Reiner A. Temporal relationship of choroidal blood flow and thickness changes during recovery from form deprivation myopia in chicks. Exp Eye Res. 2002;74(5): 561–70.
27. Hirata A, Negi A. Morphological changes of choriocapillaris in experimentally induced chick myopia. Graefes Arch Clin Exp Ophthalmol. 1998;236(2):132–7.
28. Read SA, Collins MJ, Sander BP. Human optical axial length and defocus. Invest Ophthalmol Vis Sci. 2010;51:6262–9.
29. Smith 3rd EL, Huang J, Hung LF, Blasdel TL, Humbird TL, Bockhorst KH. Hemiretinal form deprivation: evidence for local control of eye growth and refractive development in infant monkeys. Invest Ophthalmol Vis Sci. 2009;50(11):5057–69.
30. Smith 3rd EL, Hung LF, Huang J, Blasdel TL, Humbird TL, Bockhorst KH. Effects of optical defocus on refractive development in monkeys: evidence for local, regionally selective mechanisms. Invest Ophthalmol Vis Sci. 2010;51(8):3864–73.
31. Smith 3rd EL, Ramamirtham R, Qiao-Grider Y, Hung LF, Huang J, Kee CS, Coats D, Paysse E. Effects of foveal ablation on emmetropization and form-deprivation myopia. Invest Ophthalmol Vis Sci. 2007;48(9):3914–22.
32. Smith 3rd EL. Prentice Award Lecture 2010: a case for peripheral optical treatment strategies for myopia. Optom Vis Sci. 2011; 88(9):1029–44.
33. Phillips JR, McBrien NA. Form deprivation myopia: elastic properties of sclera. Ophthalmic Physiol Opt. 1995;15:357–62.
34. McBrien NA, Gentle A. Role of the sclera in the development and pathological complications of myopia. Prog Retin Eye Res. 2003;22(3):307–38.
35. McBrien NA, Cornell LM, Gentle A. Structural and ultrastructural changes to the sclera in a mammalian model of high myopia. Invest Ophthalmol Vis Sci. 2001;42(10):2179–87.
36. McBrien NA, Adams DW. A longitudinal investigation of adult-onset and adult-progression of myopia in an occupational group. Refractive and biometric findings. Invest Ophthalmol Vis Sci. 1997;38(2):321–33.
37. Saka N, Ohno-Matsui K, Shimada N, Sueyoshi S, Nagaoka N, Hayashi W, Hayashi K, Moriyama M, Kojima A, Yasuzumi K, Yoshida T, Tokoro T, Mochizuki M. Long-term changes in axial length in adult eyes with pathologic myopia. Am J Ophthalmol. 2010;150(4):562–8.e1.
38. Rose KA, Morgan IG, Smith W, Burlutsky G, Mitchell P, Saw SM. Myopia, lifestyle, and schooling in students of Chinese ethnicity in Singapore and Sydney. Arch Ophthalmol. 2008;126(4):527–30.
39. Jones LA, Sinnott LT, Mutti DO, Mitchell GL, Moeschberger ML, Zadnik K. Parental history of myopia, sports and outdoor activities, and future myopia. Invest Ophthalmol Vis Sci. 2007;48(8): 3524–32.
40. Dirani M, Tong L, Gazzard G, Zhang X, Chia A, Young TL, Rose KA, Mitchell P, Saw SM. Outdoor activity and myopia in Singapore teenage children. Br J Ophthalmol. 2009;93(8):997–1000.
41. Morgan RW, Speakman JS, Grimshaw SE. Inuit myopia: an environmentally induced "epidemic"? Can Med Assoc J. 1975;112(5): 575–7.
42. Alward WL, Bender TR, Demske JA, Hall DB. High prevalence of myopia among young adult Yupik Eskimos. Can J Ophthalmol. 1985;20(7):241–5.
43. Lv L, Zhang Z. Pattern of myopia progression in Chinese medical students: a two-year follow-up study. Graefes Arch Clin Exp Ophthalmol. 2013;251(1):163–8.
44. Mutti DO, Mitchell GL, Moeschberger ML, Jones LA, Zadnik K. Parental myopia, near work, school achievement, and children's refractive error. Invest Ophthalmol Vis Sci. 2002;43:3633–40.
45. Zylbermann R, Landau D, Berson D. The influence of study habits on myopia in Jewish teenagers. J Pediatr Ophthalmol Strabismus. 1993;30:319–22.
46. Hepsen IF, Evereklioglu C, Bayramlar H. The effect of reading and near-work on the development of myopia in emmetropic boys: a prospective, controlled, three-year follow-up study. Vision Res. 2001;41:2511–20.

47. Kinge B, Midelfart A, Jacobsen G, Rystad J. The influence of near-work on development of myopia among university students: a three-year longitudinal study among engineering students in Norway. Acta Ophthalmol Scand. 2000;78:26–9.
48. Rose KA, Morgan IG, Ip J, et al. Outdoor activity reduces the prevalence of myopia in children. Ophthalmology. 2008;115:1279–85.
49. Rucker FJ, Wallman J. Chick eyes compensate for chromatic simulations of hyperopic and myopic defocus: evidence that the eye uses longitudinal chromatic aberration to guide eye-growth. Vision Res. 2009;49(14):1775–83.
50. Young SE, Walsh FB, Knox DL. The tilted disk syndrome. Am J Ophthalmol. 1976;82:16–23.
51. Prost M, De Laey JJ. Choroidal neovascularization in tilted disc syndrome. Int Ophthalmol. 1988;12(2):131–5.
52. Quaranta M, Brindeau C, Coscas G, Soubrane G. Multiple choroidal neovascularizations at the border of a myopic posterior macular staphyloma. Graefes Arch Clin Exp Ophthalmol. 2000;238:101–3.
53. Cohen SY, Quentel G, Guiberteau B, Delahaye-Mazza C, Gaudric A. Macular serous retinal detachment caused by subretinal leakage in tilted disc syndrome. Ophthalmology. 1998;105:1831–4.
54. Cohen SY, Quentel G. Chorioretinal folds as a consequence of inferior staphyloma associated with tilted disc syndrome. Graefes Arch Clin Exp Ophthalmol. 2006;244:1536–8.
55. Becquet F, Ducournau D, Ducournau Y, Goffart Y, Spencer WH. Juxtapapillary subretinal pigment epithelial polypoid pseudocysts associated with unilateral tilted optic disc: case report with clinicopathologic correlation. Ophthalmology. 2001;108(9):1657–62.
56. Mauget-Faÿsse M, Cornut PL, Quaranta El-Maftouhi M, Leys A. Polypoidal choroidal vasculopathy in tilted disk syndrome and high myopia with staphyloma. Am J Ophthalmol. 2006;142(6):970–5.
57. Cohen SY, Quentel G. Uneven distribution of drusen in tilted disc syndrome. Retina. 2008;28(9):1361–2.
58. Vuori ML, Mäntyjärvi M. Tilted disc syndrome may mimic false visual field deterioration. Acta Ophthalmol. 2008;86(6):622–5.
59. Nakanishi H, Tsujikawa A, Gotoh N, et al. Macular complications on the border of an inferior staphyloma associated with tilted disc syndrome. Retina. 2008;28(10):1493–501.
60. Cohen SY, Dubois L, Ayrault S, Quentel G. T-shaped pigmentary changes in tilted disk syndrome. Eur J Ophthalmol. 2009;19(5):876–9.
61. Ohno-Matsui K, Shimada N, Nagaoka N, Tokoro T, Mochizuki M. Choroidal folds radiating from the edge of an inferior staphyloma in an eye with tilted disc syndrome. Jpn J Ophthalmol. 2011;55(2):171–3.
62. Maruko I, Iida T, Sugano Y, Oyamada H, Sekiryu T. Morphologic choroidal and scleral changes at the macula in tilted disc syndrome with staphyloma using optical coherence tomography. Invest Ophthalmol Vis Sci. 2011;52(12):8763–8.
63. Spaide RF, Fisher Y. Removal of adherent cortical vitreous plaques without removing the internal limiting membrane in the repair of macular detachments in highly myopic eyes. Retina. 2005;25(3):290–5.
64. Westheimer G. Entoptic visualization of Stiles-Crawford effect. An indicator of eyeball shape. Arch Ophthalmol. 1968;79(5):584–8.
65. Mäntyjärvi M, Tuppurainen K. Colour vision and dark adaptation in high myopia without central retinal degeneration. Br J Ophthalmol. 1995;79(2):105–8.
66. Mehdizadeh M, Nowroozzadeh MH. Dome-shaped macula in eyes with myopic posterior staphyloma. Am J Ophthalmol. 2008;146:478; author reply –9.
67. Imamura Y, Iida T, Maruko I, et al. Enhanced depth imaging optical coherence tomography of the sclera in dome-shaped macula. Am J Ophthalmol. 2011;151:297–302.