Ectopia lentis: contact lens fitting and diseases with this finding

Ectopia lentis: adaptação de lentes de contato e discussão das doenças relacionadas a esse achado

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KEYWORDS:

Congenital abnormalities; Contact lenses; Ectopia lentis; Lens subluxation.

ABSTRACT

Ectopia lentis is the displacement of the lens from its primary position. The lens may be completely (luxated) or partially (subluxated) dislocated. Ectopia lentis can be acquired or related to ocular and systemic diseases. Its physiopathology is related to the disturbance of zonular fibers, and lens displacement can cause many problems. This paper presents two cases of contact lens fitting in ectopia lentis and a review of diseases with this finding. Papers were extracted from the PubMed database using varying combinations of the following search terms: "Ectopia lentis", "contact lens", "Marfan syndrome", "Homocystinuria", "Weill-Marchesani syndrome", and "ocular trauma". Articles related to the identification of ectopia lentis and associated factors, such as etiology, systemic association, diagnosis, and management, were included. The reference lists of the selected articles were reviewed to obtain additional relevant articles.

PALAVRAS-CHAVE:

Anormalidades congênitas; Lentes de contato; Ectopia lentis; Subluxação do cristalino.

RESUMO

Ectopia lentis é o deslocamento da lente (cristalino) de sua posição anatômica normal. A lente pode estar completamente deslocada (luxada) ou parcialmente deslocada (sub-luxada). A ectopia lentis pode ser adquirida ou estar relacionada a doenças oculares e sistêmicas. A fisiopatologia da doença está relacionada à alteração das fibras zonulares, sendo que o deslocamento do cristalino pode causar muitos problemas. Dois casos de lentes de contato associados à ectopia lentis são apresentados, como também uma revisão para discutir as doenças relacionadas a esta síndrome. Documentos da base de dados PubMed foram extraídos usando combinações variáveis dos termos de pesquisa "ectopia lentis"; "lentes de contato"; "síndrome de Marfan"; "Homocistinúria"; "síndrome de Weill-Marchesani"; "trauma ocular". Artigos relacionados à identificação da ectopia lentis e fatores associados foram incluídos na pesquisa, tais como etiologia, associação sistêmica, diagnóstico e tratamento. As listas de referência dos artigos selecionados foram revistas a fim de se obter artigos adicionais relevantes.

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INTRODUCTION

Ectopia lentis (EL) is a condition where the lens is displaced from its anatomical position[1-3]. The lens can be luxated (dislocated) when zonular fibers rupture (total displacement). The lens may migrate to the anterior or vitreous chamber. The lens can be also subluxated (partially dislocated) when some zonular fibers rupture (or sagging). In this case, the lens remains in the posterior chamber in the pupillary area[3-5].

EL has two basic causes, i.e., hereditary or secondary. Secondary causes include trauma, high myopia, buphthalmos, anterior uveal tumors, pseudoexfoliaton syndrome, hypermature cataracts, and surgical complications[2,3,6]. Hereditary causes can be divided into causes without systemic associations (e.g., familial EL and EL et pupillae) and those with systemic associations (e.g., Marfan syndrome, homocystinuria, Weill–Marchesani syndrome, hyperlysinemia, sulfite oxidase deficiency, and Ehlers–Danlos syndrome)[2,3]. Lens displacement can lead to anisometropia, diplopia, refractive errors, and amblyopia[7-10].

Herein, we describe two unusual cases of lens displacement managed with contact lens (CL), in addition to reviewing diseases related to EL.

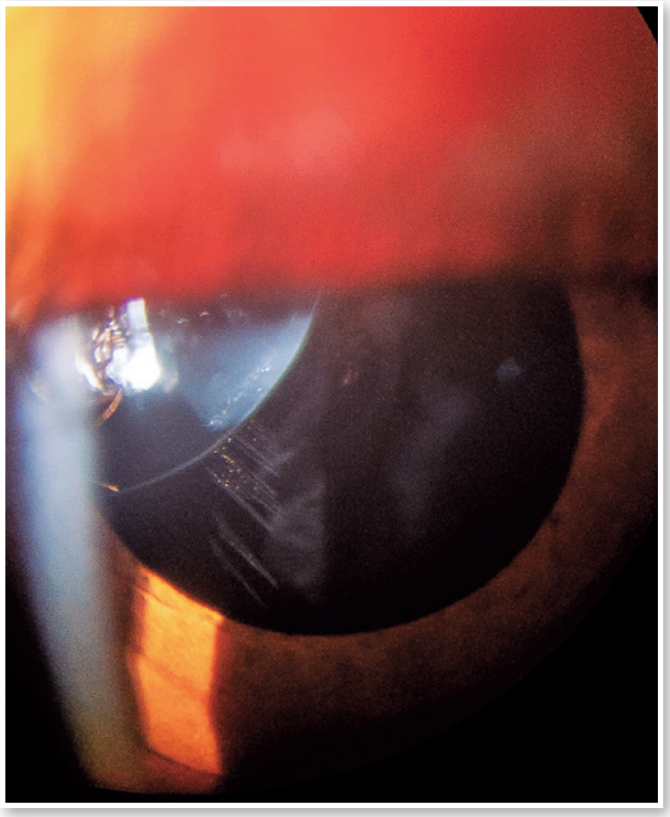

Figure 1. Biomicroscopy of the right eye.

CASES REPORT

Case 1

A.J.C.B. is a 6-year-old girl who was originally from Campinas, SP. According to her family members, she has had poor vision since birth. Her parents are first cousins. The patient did not have syndromic characteristics and had no previous trauma or family history of EL.

Ophthalmological examination:

- Refraction: OU +10,00 20/200
- Biomicroscopy: both lenses were subluxated superior temporally (Figures 1 and 2)
- Fundoscopy: attached retina, physiological cupping of the optic nerve, and preserved macula
- Corneal topography:
  OD: 46.23 x 48.76 @ 21°
  OS: 46.71 × 49.54 @ 173°
- Management: spherical rigid gas-permeable CLs (RGPCLs) were fitted
- CL parameters
  OD: base curve (BC), 46.00 D; dioptric power (DP), +20.00 D; diameter (D), 8.8mm

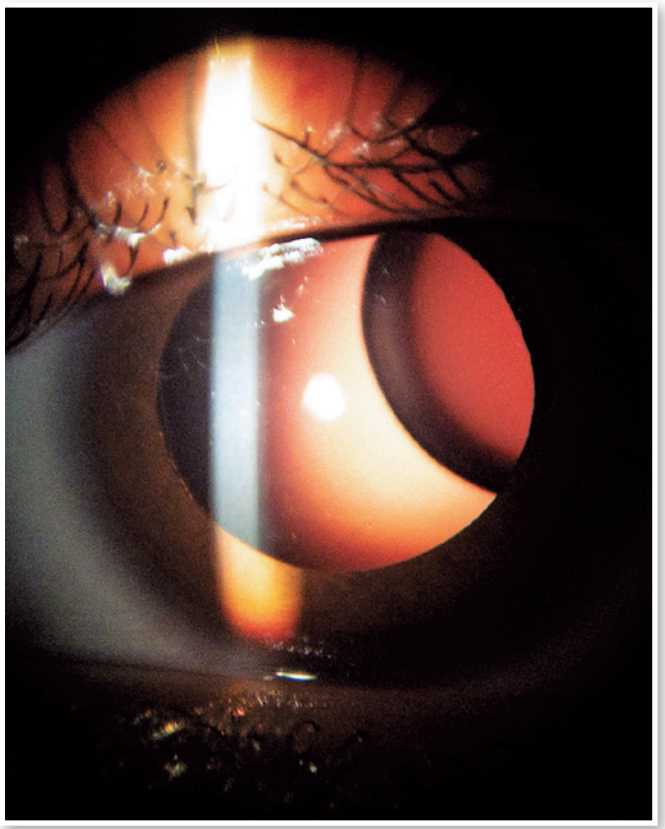

Figure 2. Biomicroscopy of the left eye.

OS: base curve (BC), 46.00 D; dioptric power (DP), +10.00 D; diameter (D), 8.8mm
Visual acuity with CL: 20/100 OU

Case 2

A.P.S. is a 57-year-old male patient who was originally from Campinas, SP. He had undergone bilateral lens extraction after spontaneous dislocation of the lenses to the anterior chamber. The patient did not have any syndromic characteristics. He had no history of trauma or family history of EL.

- Refraction
  OD: +12.50-1.50 180 20/30
  OS: + 12.00 20/30
- Biomicroscopy
  OU: aphakia and corectopia
- Corneal topography:
  OD: 42.78 × 44.21 @ 148°
  OS: 42.92 × 44.05 @ 120°
- Management: spherical RGPCLs
- CL parameters
  OD: BC, 43.00D; DP, +14.50D; diameter, 9.6mm (Figure 3)
  OS: BC, 42.00D; DP, +14.75D; diameter, 9.6mm
  Visual acuity with CL
  OD: 20/25
  OS: 20/30

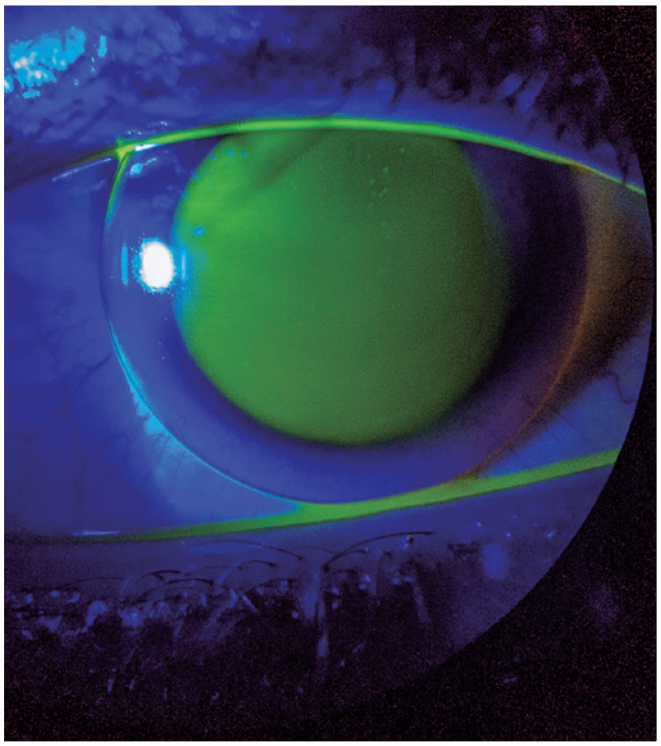

Figure 3. Right eye contact lens fitting.

Literature review

Description and etiology

Berryat was probably the first to describe the clinical findings of lens subluxation in a patient in 1749. In 1856, Karl Stellwag von Carion, a Czech ophthalmologist, introduced the term "ectopia lentis" for a patient with congenital lens dislocation. However, it was only after many years that it was associated with other eye or systemic diseases[2].

In EL, signs and symptoms depend on the extension of subluxation[11]. Low visual acuity is the main symptom. The degree of lens displacement and association with other ocular changes can lead to ametropias and/or anisometropias and low visual acuity[7,12].

Anterior displacement and rotations around the axis of the lens can lead to lenticular myopia. This can happen because of the relaxation of the zonular fibers[2,13]. If the lens edge is in front of the pupillary area, the patient may perceive optical distortions and diplopia. Bilateral cases result in quadriplopia[2,3].

EL is associated with numerous complications that worsen the visual prognosis, such as cataracts, secondary glaucoma (due to anterior chamber angle malformation), and retinal detachment[10,12,14].

The EL classification system classifies EL according to the movement of the lens (displacement or subluxation): displacement is defined as complete movement, which can be anterior or posterior, and subluxation is defined as the movement of the lens in the coronal plane behind the iris. This classification system is relevant for standardizing the evaluation and detailing the movements of the lens[2,3].

EL without systemic associations

Simple EL or familial EL

Simple EL, or familial EL, is an autosomal dominant disease without associated systemic manifestations. An autosomal recessive form has also been reported[15-18]. It can present as a congenital clinical condition or occur spontaneously with a late onset, the latter being the most common form, occurring between the age of 20 and 65 years[2,19].

Mutations occur in the *fibrillin-1* gene (*FBN1*) mapped to chromosome 15q21[3]. There may be an overlap between the genetics of simple EL and EL associated systemic diseases because mutations in the *FBN1* gene can also occur in Marfan syndrome[9,20,21].

In this condition, irregularity and degeneration of the zonular fibers occur, which causes lens subluxation. It mainly manifests as bilateral, symmetrical,

upward, and temporal lens displacement[5,17,20]. However, in some cases, the degree of lens displacement can vary between the two eyes and can be asymmetric.[2,3] Possible complications are retinal detachment, secondary glaucoma, and total lens displacement[20,22]. These patients may progress with cardiovascular changes, and they must have long-term cardiologic follow-up because of its relationship with the Marfan syndrome gene[20,23].

EL et pupillae

This is a rare congenital disease, a unique condition that is characterized by concomitant ectopy of the pupil and lens[10]. It is autosomal recessive and may be related to a mutation in the *ADAMTSL4* gene on chromosome 1q21[24-26].

The diagnosis is essentially clinical and important for risk assessment, prognosis, and treatment[10,27]. In most cases, both eyes are involved, which may be symmetrical or not, and the pupils are retracted, oval, or elliptical and displaced in the direction opposite to the EL[28,29]. However, 40% of the patients present only poor pupillary dilation[18,28,30]. Eye abnormalities, syndromes, and metabolic disorders are part of the differential diagnosis of this disease[24,27]. The pathogenesis is still unknown. However, theories have advocated the occurrence of mesodermal and neuroectodermal changes[17,26,31]. With these changes, a persistent and abnormal vascularized tissue is formed, which anastomoses with the hyaloid system and generates pupillary ectopia and malformation of the zonule in the corresponding area[10,17,26]. Cases with microspherophakia and transillumination of the iris have also been documented[10,25,26].

Uveitis is relatively common, and the main mechanisms are direct contact of the displaced lens with the ciliary body or the posterior face of the iris, leading to acute iridocyclitis, and increased permeability or rupture of the anterior lens capsule, with protein leakage, causing phacolytic uveitis[24,30].

EL associated with systemic diseases

Marfan syndrome

This syndrome was described in 1896 by the French pediatrician Antoine Bernard-Jean Marfan. It is an autosomal dominant systemic disorder of the connective tissue caused by a mutation in the *FBN1* gene on chromosome 15q21[2,3]. The manifestations include cardiovascular, ocular, and skeletal involvement. The disease has no predilection for sex, affecting both men and women equally[32-36,38].

Skeletal clinical findings include excessive growth of the long bones, which generates thoracic deformity, disproportionate limbs, scoliosis, arachnodactyly, among others[36,37]. Myalgia, fatigue, and muscle hypoplasia are usually present[2,9].

Patients with cardiovascular involvement present mitral valve dysfunction, with prolapse, insufficiency, and calcification. Cardiac manifestations are the main causes of morbidity and mortality in Marfan syndrome. Aortic dilation, dissection, aneurysm, and even rupture of the aortic root can occur[9,38,39].

EL is the most common ocular manifestation. It affects approximately 80% of patients, with 70% of cases aged up to 6 years[3,9]. The subluxation is mainly superior temporal or superior nasal, the lens border is irregular, and zonular fibers are long and can be either completely broken or retracted[14,22,32]. Coloboma of the lens may occur; however, it is a less common ocular manifestation. It is characterized by a notch at the equator of the lens, with absent or underdeveloped zonules. It is also usually monocular and may or may not be associated with other systemic abnormalities[34]. Other ophthalmological findings may include exophthalmos caused by decreased retro-orbital adipose tissue, megalocornea, iris transillumination, high myopia, retinal detachment, cataracts, or glaucoma[40,41].

In Marfan syndrome, EL is classified into five stages, which assess the positive predictive value of EL according to the eye examination: anteroposterior displacement (stage 1, minimal displacement), anteroposterior displacement and superior displacement (stage 2), subluxation and elongation of the lower zonular fibers (stage 3), subluxation and breaking of some zonular fibers (stage 4), and dislocation (stage 5). From stage 2, EL has a high positive predictive value for the clinical diagnosis of Marfan syndrome[37].

In 2010, the Ghent nosology criteria, used for the diagnosis of Marfan syndrome, were updated. Patients with EL and a *FBN1* mutation are now categorically diagnosed with MFS if their mutation has previously been described with aortic dilation/dissection[33].

The increase in life expectancy in Marfan syndrome is related to the early approach to cardiovascular complications[39].

Homocystinuria

This disease is characterized by the deficiency of cystathionine β-synthase and methionine synthase, enzymes responsible for homocysteine metabolism, and it is caused by mutations in the *CBS* gene on chromosome 21q22.3[3]. Its inheritance is autosomal recessive[42].

Homocysteine is detected in urine tests but may be present in conditions other than homocystinuria. Urine chromatography or high-voltage electrophoresis can confirm the diagnosis[2].

Homocysteine accumulation generates cardiovascular, skeletal, and ocular manifestations. The main manifestations are progressive cognitive delay and inferonasal lens subluxation (which is progressive and present in approximately 90% of the patients and results from degenerative changes in the zonular fibers)[42,43].

Lenticular myopia is also common and usually precedes the EL[2,14]. These patients have appearance similar to patients with Marfan syndrome and may have early thromboembolic events and skeletal abnormalities, including osteoporosis, genu valgum, and thinning and elongation of long bones. The main mortality from the syndrome is related to thrombotic vascular occlusions[42].

The patient must maintain a diet low in methionine and rich in cysteine, with supplementation of pyridoxine (vitamin B6), which can prevent or delay mental retardation and lens subluxation[11,43].

Weill–Marchesani syndrome

This syndrome was described by Georges Weill in 1932 and delineated by Oswald Marchesani 7 years later[3]. It is a rare syndrome caused by a genetic mutation in ADAMTS or FBN1 proteins, which generate a connective tissue disorder and can have an autosomal dominant and recessive inheritance with occasional brachymorphism in heterozygotes[2,3,44].

This syndrome is clinically characterized by microspherophakia (in some cases, the lens has 50% of its normal volume), short stature, brachydactyly, thick skin, stiff joints, and occasional heart defects.[3] Osteoporosis was also been described in this syndrome[45].

Microspherophakia is a primary ocular manifestation and is considered a diagnostic criterion for this syndrome. It is characterized by small, spherical lens, with lesser, elongated, and relaxed zonular fibers around its equator[13]. Lenticular myopia and lower lens displacement can also be present. The prevalence of associated glaucoma is also high because of lens displacement and contact with the iris, which can cause visual loss[3,44]. Other ocular abnormalities associated with Weill-Marchesani syndrome are megalocornea, nonspecific chorioretinal degenerations, and scleral staphyloma[2,3].

Hyperlysinemia

This is a rare systemic disease caused by an innate error in the metabolism of lysine, an essential amino acid. It is caused by the mutation of the *AASS* gene on chromosome 7q31.3s, is an autosomal recessive disease, and has a high association with inbreeding[2,3].

Clinically, patients have a cognitive delay and muscle hypotonia and may present with EL, recurrent vomiting, lethargy, diarrhea, and developmental delay[3,19].

When EL occurs, it presents as bilateral subluxation and may be associated with paresis of the extraocular muscles[2].

Sulfite oxidase deficiency

The first case of this syndrome was described in 1967 by Irrevere. It is a rare disease of autosomal dominant cause, and the gene implicated is the human sulfite oxidase gene (*SUOX*) located in chromosome 12q13.13. A change in the metabolism of methionine and cysteine generates increased urinary excretion of S-sulfocysteine, taurine, sulfite, and thiosulfate[2,3].

The central nervous system is involved because of the accumulation of sulfite in the brain, generating severe and generalized losses of neurons, myelin, and axons, with special glial proliferation. Therefore, the patient may present changes in muscle tone, refractory seizures, dystonia, and cognitive delay[2,19]. EL has been reported in 53% of cases, being diagnosed between 3 months and 3 years of age. The pathogenesis of EL in sulfite oxidase deficiency is still unclear[3].

Ehlers–Danlos syndrome

This syndrome was named after Edvard Ehlers, a Danish dermatologist, and Henri-Alexandre Danlos, a French dermatologist, who recognized it, respectively, in 1901 and 1908[46].

Being a rare disease, 50% of its classic types are caused by mutations in *COL5A1* (chromosome 9q34.3) and *COL5A2* (chromosome 2q32.2) genes

that encode type V collagen alpha 1 and alpha 2 chains. It has no racial predisposition and occurs in 1 of 5,000 live births[3].

It is characterized by generalized fragility of soft connective tissues, cutaneous hyperextensibility, delayed healing, joint hypermobility, and bruising. EL occurs occasionally, and other ocular manifestations may be present, such as scleral fragility, keratoconus, and myopia[3].

Diagnosis

When attending to a patient with EL, the physician must pay attention to the patient's age and personal and family history of comorbidities. During physical examination, syndromic characteristics and skeletal, cardiovascular, and eye alterations should be sought[14,30].

During the ophthalmological evaluation, visual acuity, refraction, and comprehensive ocular examination must be performed. When the patient is in mydriasis, sometimes the edge of the lens and the zonular fibers can be observed.

Every patient with suspected hereditary EL should undergo a systemic evaluation with metabolic screening, echocardiography, and musculoskeletal examination[7,19,39].

Gonioscopy must be performed in all patients because angular narrowing and changes in the sinus of the chamber may occur. Iridocorneal changes, such as megalocornea and corectopia, should be also investigated[27,30].

Imaging examinations can be used for diagnostic assistance, such as ocular ultrasonography, computed tomography, and magnetic resonance imaging. Ultrasonography is the first option in terms of availability and can identify complications such as retinal detachment[47].

Management

Clinical follow-up of the patient and optical correction with glasses or CLs are still the classical approach for EL, especially in cases with mild and stable subluxation and without complications[9,48-50].

Optical correction is useful for the prevention of amblyopia, especially in patients with low and stable refractive errors[14,30].

Contact lenses have several advantages over glasses, such as fewer optical aberrations, elimination of peripheral distortions, prismatic effects, among others. Rigid gas-permeable lenses are the best option in terms of visual acuity; however, some authors recommend silicone-made CLs as the first choice because of their greater oxygen permeability, thermal conductivity, ease of placement, and relative safety. Thus, the choice of the lens type depends on the child's adaptive issues and family compliance[51].

Surgical treatment may be necessary in cases of high lenticular astigmatism, rotation of the lens on its axis, lens mobility with unstable refractions, and lens edge located on the visual axis[52,53]. In these patients, each case must be individualized, as there are high risks inherent to surgical procedures. In general, the lower the degree of subluxation, the safer the surgical procedure[54-56].

Several surgical techniques are available, and each indication depends on the patient, eye conditions, disease that caused lens displacement, and surgeon's skill, experience, and preference. Although outcomes of surgical management are still limited, the margin of treatment success increased owing to advances in surgical techniques and materials[55,56].

Some of the most widely used surgical techniques are extracapsular facectomy or phacoemulsification with or without implantation of IOL in the ciliary sulcus or capsular sac, and phacofragmentation with posterior vitrectomy via the pars plana with IOL fixation in the sclera or iris. For better visualization during surgery, an endocapsular expander ring should be used, especially in extracapsular facectomy or phacoemulsification[52,57].

The most common surgical indication in EL is when the lens edge reaches the visual axis with extensive disinsection or a lack of capsular support[50,58]. In these cases, closed lensectomy–vitrectomy is performed via the pars plana or limbus. Aphakia must be corrected with CLs, glasses, or IOL implants to avoid amblyopia[59,60]. Correction with an IOL implant requires a long-term follow-up because of the risk of suture rupture and displacement of the IOL over time. In the absence of adequate capsular support, scleral fixation of the foldable IOL in the posterior chamber is the current procedure of choice, and the suture fixation technique showed greater advantages with fewer complications[59-62].

When the lens is displaced by >180°, the use of Cionni's modified capsular tension ring allows the best surgical visualization and fixation of the capsular bag in the sclera. It also maintains the integrity of the capsular bag in extensive zonular lesions, preventing the collapse of the capsular bag after lens removal and

offers safety during phacoemulsification and implantation of the IOL, keeping the capsule stable[63-66].

In Marfan syndrome-associated EL, if the patient does not obtain good visual acuity following clinical correction or presents complications, lensectomy with or without secondary IOL implantation is indicated[40,53,65].

In EL et pupillae, the main approach is the clinical management. Early optical correction should be performed to avoid amblyopia. In patients who have an off-axis pupil, optic or mydriatic iridectomy can be performed[10,12].

In homocystinuria-induced EL, surgical treatment should be considered, especially in cases of lens displacing to the anterior chamber or glaucoma caused by pupillary block[67].

Spontaneous displacement of the lens to the anterior chamber, as in one of the cases reported in the present article, is an ophthalmological urgency because of the risk of corneal decompensation and glaucoma[68]. Postoperative complications, such as vitreous loss and retinal detachment, can be avoided with lensectomy and vitrectomy via the pars plana[7,11].

Genetic counseling should be always considered. To avoid trauma, patients should avoid contact sports or maneuvers that may increase the intraocular pressure because ocular complications are more frequent in these groups than in the normal population[14].

EL can be associated with several systemic or isolated diseases. Anamnesis and clinical examination are pivotal for diagnosis. During the clinical evaluation, the ophthalmologist should try to recognize syndromic characteristics in every patient because EL can be a feature of those diseases.

Currently, clinical treatment is the first option because surgical treatment can present several complications. In the first case, considering the patient's age, surgery would predispose the child to complications, such as aphakia, uveitis, anterior and posterior synechiae, secondary glaucoma, and retinal detachment. The use of CL is an alternative to postpone surgery until they reach an age when complication rates are lower. CLs can provide satisfactory visual acuity to develop the visual pathways and prevent amblyopia. In the second case, CL adaptation was also useful, as it provided a good visual acuity and gave the patient the best aesthetics. The management of EL remains a challenge, especially in children.

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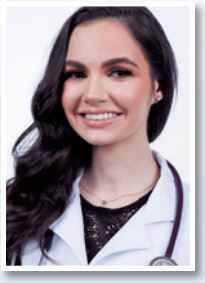

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