ABSTRACT
Objective: To present a classification system to grade \textit{ectopia lentis} and to assess its usefulness as a predictor for surgical outcomes. Methods: Fifty-one eyes of 28 patients with either simple (19 patients) or Marfan syndrome-associated \textit{ectopia lentis} (nine patients) with variable degrees of subluxation were operated on. Lens subluxation intensity was graded according to the lens subluxation grading system (LSGS) from grade 1 (lens on the whole pupillary area) up to grade 4 (lens absent from the pupillary area). Thirty eyes underwent cataract extraction ("dry" aspiration) with endocapsular ring and in-the-bag intraocular lens (IOL) implantation. Twenty-one eyes underwent cataract extraction ("dry" aspiration) with scleral fixation of the IOL. The predictive value of the LSGS was assessed by analyzing the post-operative outcomes, including visual acuity (VA), endothelial cell loss, and complications for each grade on the grading system. Results: Patients were classified into grade 1 (19.6%), grade 2 (51%) and grade 3 (29.4%). Post-operative VA was lower for eyes with larger degrees of subluxation. The higher the subluxation grade, the higher the endothelial cell loss, as well as, the frequency of vitreous loss and surgical time. Higher subluxation grades prevented optimal surgical outcomes with endocapsular ring and in-the-bag IOL implantation. Conclusions: The LSGS provides an estimate of the surgical success of \textit{ectopia lentis}. Adequate standardization of lens subluxation is crucial for understanding studies dealing with the surgical correction of this disorder.

Keywords: Ectopia lentis/surgery; Marfan syndrome; Lens subluxation; Lens implantation, intraocular

INTRODUCTION
\textit{Ectopia lentis} is a lenticular dystopia, either congenital or acquired in origin, secondary to trauma or spontaneous. According to the amount of zonular support, the dystopia...
can be classified into subluxation, when the lens partially remains in the pupillary area at the posterior chamber, or luxation, when it can migrate to the anterior chamber or vitreous cavity as a result of complete rupture of zonule fibers\(^1\).

Spontaneous lens subluxation can occur as result of an anomalous zonular development, making it short in length and spherical in shape. It is frequently a family condition and the zonular fibers can be very elongated and remain adhered to the lens\(^2,3\). The lens can become opaque and cause increased intraocular pressure and its extraction is recommended\(^2,5\).

The lens equator can be seen in the pupillary area, even without mydriasis. In this situation monocular dyopia or binocular quadruplication can occur\(^5\). The zonular fibers can vary in shape and grouping on biomicroscopy. Zonular weakness causes the lens to become spherical in shape, thus leading to severe myopia. The lens are usually shorter; they can remain transparent and, occasionally, they are colobomatous\(^5\).

The displacement of the lens usually results in optical complications. In addition, glaucoma, uveitis, retinal detachment and cataract can occur\(^6\).

The treatment of *ectopia lentis* poses a challenge to clinicians. Not infrequently, a multidisciplinary team is involved for caring for systemic abnormalities and genetic counseling\(^3\). The therapeutic approach should be individualized and the goal should be to provide good visual acuity (VA)\(^3\).

Optical correction with glasses or contact lenses is the first choice for many patients. The use of mydriatic or miotic agents can help to improve vision\(^6\). The surgical treatment of *ectopia lentis* is controversial. While some authors delay lens extraction for fear of intraoperative and post-operative complications, others believe that subluxation itself is an indication for surgery\(^7,8\).

Careful assessment of specific indications for surgery is suggested\(^9,4\). Continuous development of ocular instruments and improvement of surgical techniques have stimulated many people to choose the surgical management to improve VA in this patients\(^9,10,15,19-25\). Many authors have proved the efficacy and safety of different techniques for the surgical correction of *ectopia lentis*\(^12,15,19-25\).

Although the majority of surgeons are unanimous on the indication for early surgery on severe subluxation cases, none has clearly established a lens subluxation severity graduation.

### OBJECTIVE

The purpose of this study is to propose a classification system for grading lens subluxation and to evaluate its predictive value for surgical outcomes.

### METHODS

#### Sample and selection criteria

This prospective study was approved by the Ethics Committee from the two Institutions involved (Department of Ophthalmology of Santa Casa de São Paulo and Faculdade de Medicina da Universidade de São Paulo) and the procedures were in agreement with the tenets of the Declaration of Helsinki.

Patients were enrolled in the study from March, 2000 to June, 2002 based on the presence of lens subluxation on examination. Exclusion criteria included lens luxation to the anterior or posterior chamber, history of ocular trauma, corneal opacity precluding anterior segment examination, glaucoma, uveitis, retinal abnormalities compromising visual recovery or posing risk for retinal detachment, previous ocular surgery, any medical condition preventing patients from undergoing general anesthesia and patient unwillingness. All subjects underwent a complete eye examination including refraction, biomicroscopy, aplanation tonometry, indirect ophthalmoscopy, A-scan, and endothelial cells count. Fifty-one eyes from 28 patients were enrolled in the study. Nineteen patients had simple *ectopia lentis* (SEL) and nine had Marfan syndrome-associated *ectopia lentis* (MS). Sixteen patients were male and 12 were female. Mean age was 15.9 ± 8.5 years (range, 5.4 to 37.6 years).

#### Lens subluxation grading system

In order to assess subluxation severity, it was proposed a classification system based on the lens displacement in relation to the undilated pupil. The patient’s eye was assessed at the slit-lamp with 10x magnification, broad slit for diffuse illumination in order to achieve a wide panoramic view. A narrow slit with 45° illumination for locating the lens was used when necessary. Assessment was completed with the red field illumination and the result expressed into one of four grades (Figure 1): grade 1, lens seen on the pupillary area; grade 2; lens seen on 2/3 of the pupillary area; grade 3, lens seen on 1/2 of the pupillary area or grade 4, lens absent from the pupillary area.

#### Surgical technique

Surgery was indicated when the patients VA prevented them from doing their daily activities, i.e., job, study, domestic tasks, driving, leisure, etc. The surgeries were performed by only one surgeon (MW). Twenty-three patients (82.1%) had bilateral, sequential surgery; 24 eyes were right eyes (47.1%) and 27 were left eyes (52.9%).
The procedure was initiated with the creation of a scleral frown incision (5 mm) using a crescent knife (Alcon Surgical, Fort-Worth, Texas) and a temporal or nasal corneal incision (1 mm). The anterior chamber was filled with a viscoelastic agent and capsulorrhexis was done with an Utrata forceps. At this point, an endocapsular ring (CTR -10, Mediphacos, Belo Horizonte – Minas Gerais, Brazil) was inserted. In the eyes in which the capsulorrhexis was suboptimal, the endocapsular ring was not implanted.

Following, it would be the hydrodissection, the lens content was removed with “dry” aspiration by means of a canula and syringe and viscoelastics were used to deepen the anterior chamber as needed. A polymethylmethacrylate (PMMA) intraocular lens (IOL) was placed in the bag and the scleral ring was sutured at the sclera. In eyes in which an endocapsular ring was not inserted, scleral fixation of the PMMA IOL was done with 10-0 polypropylene sutures. The scleral wound was sutured with one 10-0 nylon suture and a sub-conjunctival injection of gentamycin 1 cc and dexamethasone 1 cc was given at the inferior fornix.

Statistical analysis
It was used the $\chi^2$ test to compare subluxation grades between eyes with SEL and MS. The improvement of VA at each visit was analyzed with ANOVA. Unpaired t-Student test and Mann-Whitney U test were used to compare axial length between SEL and MS eyes; pre and post-operative endothelial cell loss between the two surgical techniques; post-operative VA (at month 6) and duration of surgery between eyes with intraoperative vitreous loss and eyes with no vitreous loss. Pre and post-operative endothelial cell loss for the 51 eyes were compared with paired t-Student test. ANOVA and Kruskal-Wallis test were used to compare axial length and endothelial cell loss among each LSGS group. A p value of less than 0.05 was considered to be of statistical significance.

RESULTS
Preoperative lens subluxation was classified as grade 1 in 10 eyes (19.6%), grade 2 in 26 eyes (51%), and grade 3 in 15 eyes (29.4%) according to LSGS. No patient had grade 4. These figures were similar for SEL and MS patients ($\chi^2 = 1.63; p = 0.443$).

Mean preoperative uncorrected VA was not different for each LSGS group (Table 1), although lower LSGS grades had better VA. As to the post-operative uncorrected VA, eyes with grade 3 presented with lower VA at each time point throughout the follow-up as compared to grades 1 and 2 (Figure 2). However, at the last follow-up, eyes with LSGS grades 1 and 2 had better VA as compared to grade 3 (Table 2).

The mean axial length for the 51 eyes was 24.6 ± 2.0 mm and eyes with MS presented longer measurements (25.5 mm) as compared to those with SEL (24.2 mm) (t = 2.09; p = 0.04; U = 202.5; p = 0.08). Analysis of axial length stratified per LSGS grades showed no difference (F = 1.73; p = 0.188; H = 4.42; p = 0.109).

Preoperative endothelial cell count was 3,497.2 cells/mm$^2$ and post-operative count was 3,154.2 cells/mm$^2$. This difference (343 cells/mm$^2$; 9.8%) was statistically significant (t = 8.41; p < 0.0000001). Difference between SEL (10.3%) and MS (8.7%) was not significant. The endothelial cell loss increased proportional to higher LSGS grades (grade 1, 6.7% loss; grade 2, 9.5% loss; and grade 3, 12.3% loss); these differences, however, failed to reach statistical significance (F = 1.47; p = 0.240; H = 2.59; p = 0.247).
When compared as to the surgical technique, endothelial cell loss was 12.8% in eyes that underwent cataract extraction with scleral fixation of the IOL and 7.6% in eyes that had cataract extraction with endocapsular ring and in-the-bag IOL implantation (t = 2.29; p = 0.02; U = 224; p = 0.08).

Intraoperative vitreous loss was recorded in five eyes (9.8%). Vitreous loss occurred in one eye with LSGS and grade 2 and 4 eyes with LSGS grade 3. Uncorrected VA at the last follow-up was 0.21 for eyes with vitreous loss as compared to 0.42 for the remaining 46 eyes (t = 2.24; p = 0.02; U = 224; p = 0.08). Best corrected VA was 0.37 for eyes with vitreous loss and 0.65 for the remaining 46 eyes (t = 2.36; p = 0.02; U = 53; p = 0.04). Mean surgical time was 93 minutes for the five eyes with vitreous loss, 34.1% longer than the 69.3 minutes for the 46 eyes with no vitreous loss (t = 3.47; p = 0.001; U = 48; p = 0.03).

Complications were recorded in three eyes (5.9%) and included secondary glaucoma in two eyes (3.9%) well controlled with anti-glaucomatous topical medication. Post-operative displacement of the IOL occurred in one eye (2.0%), which was successfully replaced later on.

**DISCUSSION**

Fast recovery and adequate optical correction of refractive errors in patients with *ectopia lentis* are crucial for preventing the development of amblyopia in infants. When the subluxation is small and stable, the refractive error can be small and easily corrected with eyeglasses or contact lenses. When no complication is evident, patients should not be operated on (1,4,6,9,19,26-27).

On the other hand, lens dislocation can cause optical defects which cannot be easily corrected by means of glasses and contact lenses. High astigmatism can be induced by the lens equator placed in the median pupillary portion or due to irregular shape and rotation over its axis as a result of partial zonular rupture. Severe lenticular myopia can result from partial absence of zonules (28).

The most common cause of surgical indication to correct *ectopia lentis* is the pupil bisection through the lens equator, in the visual axis. Even though visual correction for both the phakic or aphakic pupil is possible, the resulting vision might not be adequate (6). Additionally, the lens can be so unstable that refractive errors change continuously and, occasionally, complete luxation may occur.
When an acceptable VA for engaging in daily activities is not achieved, after careful refraction or when imminent signs of complications develop, surgery must be considered.

In this study, surgery was indicated when the best corrected VA was equal to or less than 20/50 or when the patient’s quality of vision was not appropriate for them to perform daily activities\(^{10-18,29-36}\).

In this paper it was decided to include both eyes of the same patient whenever the need for surgical intervention was mandatory for both. This was done considering that lens subluxation is a relatively rare condition and, by using only one eye per patient, the sample size would be reduced. A sub-analysis per group (between SEL and MS) was not carried out, since the small number of patients in each group would fail to reach a high statistical power in order to draw definite conclusions. Additionally, regardless of the cause for lens subluxation, the clinical picture and prognosis, as well as the intervention, are the same for either case.

The choice for the surgical technique was based on the safety of the procedure, short learning curve, adequacy for the complexity of the condition and dispensability of expensive pieces of equipment and technology. In this regard, the surgical technique used in these groups of patients seemed to be a good choice. In some eyes, the capsulorrhexis was not optimal and the removal of the lens content with “dry” aspiration and scleral fixation of the IOL in the posterior chamber was an alternative approach\(^{37}\).

 Previous studies have shown the efficacy of IOL implantation using the endocapsular ring for scleral fixation in the management of zonular dialysis in cataract surgery\(^{11-13,38-39}\).

It was observed that endocapsular ring implantation was feasible in eyes with less extensive lens subluxation (LSGS grade 1 and 2) in which capsulorrhexis was optimally performed. These patients were, indeed, those with better preoperative VA and less amblyopia.

The patient’s distribution was homogeneous in terms of age, gender, ethnicity, eye to be operated on, diagnosis and LSGS grades. Thus, the surgical outcomes of the study are reliable and free from possible selection bias.

VA improved steadily after surgery, especially after post-operative day 9 up to day 90 (three months). In the first post-operative days, VA is compromised by a slight corneal edema and, occasionally, by cystoid macular edema, which can resolve spontaneously\(^{26}\).

The improvement in VA was related to the LSGS grade. The present results have shown that the final VA was better in eyes with grade 1 than in those with grades 2 and 3. Eyes with higher subluxation grades demand more complex optical correction and have greater chances of developing amblyopia. The uncorrected VA was related to the surgical technique. Eyes undergoing cataract extraction and endocapsular ring had better final VA than those undergoing cataract extraction and scleral fixation. On the other hand, the cause of ectopia lentis (simple or Marfan-associated) was related to post-operative best corrected VA.

Ectopia lentis can worsen in time, so that, subluxation grades can increase\(^{11,12,19}\). Our study has shown the predictive value of the LSGS in the surgical outcomes. If worsening of the lens subluxation occurs, we believe that early surgical intervention can bring better visual results, thus decreasing the chances of amblyopia in young children\(^{28}\).

Mean uncorrected and best corrected VA were 50% lower in eyes with vitreous loss. These eyes (9.8%) had LSGS grades 2 (3.9%) and 3 (26.7%), an indication that vitreous loss is more frequent in eyes with more advanced lens subluxation. This situation was found in these eyes. Eyes with LSGS grades 2 and 3 are more likely to have intraoperative vitreous loss; eyes with vitreous loss have little improvement in VA; finally, eyes with little improvement in VA are those with higher lens subluxation grades from the start. In addition, eyes with more intense lens subluxation are prone to have more intraoperative complications and vitreous loss, demanding extra time for surgery (as reported in the cases of the present article).

Endothelial cell count decreased by 9.8% on average and eyes that underwent cataract extraction and scleral fixation of the IOL had higher loss. This technique was employed in eyes with higher LSGS grades, since optimal capsulorrhexis could not be achieved. Aspiration of lens content without the endocapsular ring had to be done more repeatedly and the suturing of the IOL caused the procedure to be more traumatic, leading to additional loss of endothelial cells.

When comparing post-operative VA with axial length, the present results showed that eyes with longer axial length presented worse VA, possibly due to preoperative amblyopia. In addition, eyes with longer axial length had higher grades of lens subluxation. We believe this is a casual relationship, i.e., subluxation increase as a proportion of the increase in the axial length. Surgery in eyes with longer axial length is more difficult as a result of higher subluxation grades.

It was recorded very few complications, namely, glaucoma in two eyes (one eye with SLGS grade 2 and one with grade 3) and subluxation of the IOL in another. Possible causes for glaucoma in these eyes would include trabecular inflammation, obstruction of
conventional outflow by red blood cells, viscoelastics or cortical debris, and distortion of the anterior chamber angle\(^1\). The two eyes with glaucoma were satisfactorily controlled on medical therapy.

The proper implantation of an IOL is mandatory, in order to achieve satisfactory VA. Optimal capsular support for adequate placement of the IOL is an exception in eyes with lens subluxation. In our series, we used the endocapsular ring prior to IOL implantation whenever possible, or the IOL would be sutured to the sclera in order to have a finely placed implant. Nevertheless, four eyes presented with minor displacement of the IOL optic, which compromised either VA or the patient’s satisfaction. One eye that had scleral fixation of the IOL underwent a second procedure for IOL resuturing. This patient denied any trauma and the reason for the IOL displacement remained unknown.

Post-operative IOL eccentricity was proportional to preoperative lens subluxation. No eye with LSGS grade 1 presented with post-operative IOL displacement, whereas two eyes with LSGS grade 2 and two eyes with LSGS grade 3 presented with slight IOL dislocation. The majority of eyes with LSGS grade 1 had endocapsular ring implantation and it is believed that this technique enhances the chances of having a properly placed IOL in the post-operative period.

**CONCLUSIONS**

Adequate classification on lens subluxation, as the one proposed here (SLGS), is helpful in predicting post-operative outcomes. The lower the amount of subluxation (SLGS grade 1 and 2), the better the visual results and the patient’s satisfaction, not to mention that the surgical procedure is carried out more safely.

**REFERENCES**


