„CAROL DAVILA”

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The use of Cionni rings in lens subluxation in Marfan syndrome

PhD THESIS SUMMARY

PhD supervisor:

PROF. UNIV. DR. CĂLIN PETRU TĂTARU

PhD student:

ANCA CRISTINA PUȘCAȘU (DOGĂROIU)

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Introduction

Marfan syndrome, first described by the French pediatrician Antoine Marfan in 1896 (1), is a connective tissue disorder caused by mutations in the FBN1 gene, which encodes the protein fibrillin-1. Mutations in this gene affect the production and secretion of fibrillin-1, the main component of the extracellular matrix, resulting in varied clinical manifestations involving multiple body systems: musculoskeletal, cardiovascular, pulmonary, integumentary, and ocular.

Ocular disorders in Marfan syndrome, essential for diagnosis, include ectopia lentis, myopia, and astigmatism, with ectopia lentis present in approximately 60% of cases (2), significantly affecting visual acuity. Over time, numerous surgical techniques have been developed to treat ectopia lentis, considering that this condition usually occurs before the age of 10 (3,4).

Currently, there is no consensus on the ideal age for surgical intervention or the optimal technique. Preserving the capsular bag and strengthening the zonular support using Cionni tension rings, anchored to the sclera, is a laborious method that requires skills and experience from the surgeon. If the capsular bag cannot be preserved, other techniques may be applied, varying in complexity.

This study retrospectively analyzes the records and video recordings of surgeries performed on patients with ectopia lentis associated with Marfan syndrome, conducted by the same surgeon, comparing four surgical techniques used. The most common technique involves implanting an artificial lens in a capsular bag stabilized with a modified Cionni tension ring.

The hypothesis is that the use of Cionni tension rings is a reproducible technique that allows the implantation of a suitable, centered, and stable lens over time, providing superior postoperative visual outcomes compared to other techniques.

The study objectives include evaluating the surgical benefits (visual acuity, stability, reproducibility) brought by Cionni tension rings, the frequency of use of Cionni rings, the rate and type of complications, evaluation and prevention of surgical incidents and difficulties, the influence of patient age on the choice of surgical technique, and the impact of the surgical technique on the selection of artificial lens design.

The study's limitations include the relatively small number of subjects enrolled (explained by the fact that Marfan syndrome is a rare pathology), short-term postoperative follow-up, lack of biometric data, variations in the surgeon's experience, uneven distribution

of subjects, and the absence of a control group.

General Part

1. Lens subluxation (ectopia lentis) in Marfan syndrome

1.1. History of Marfan syndrome and ectopia lentis surgery

Marfan syndrome was first described in 1896 by the French pediatrician Antoine Marfan, who observed unusual physical traits in a five-and-a-half-year-old girl (1): long and thin limbs, underdeveloped muscles, joint contractures, and delay in locomotor function (5). Initially, Marfan called this condition dolichostenomelia, later renamed arachnodactyly (spider fingers) by Achard (6). In 1924, Ormond and Williams first reported the correlation between Marfan syndrome and ectopia lentis (5–8), observing iridodonesis and lens subluxation in about half of the patients (6,8). In 1935, Burch (6) expanded the clinical manifestations of Marfan syndrome, including long limbs, a narrow face, dolichocephaly, chest and spine deformities, flat feet, joint hypermobility, and cardiac and pulmonary abnormalities. Additional ocular manifestations included iridodonesis, myopia, nystagmus, and divergent strabismus (6).

Historical surgical techniques for correcting ectopia lentis

Ectopia lentis surgery has been much debated over time, with discussions about the necessity of intervention, the optimal timing for surgery, and the best surgical techniques (9–12). Two categories of approaches have been described: interventions on the iris (iridodesis and iridectomy - (13,14)) and interventions on the lens (couching, lens discission, lens extraction with forceps, electrodiafaco and cryoextraction, extracapsular techniques - (13,15–20)).

1.2. Anatomy, physiology, and pathophysiology elements of the zonular apparatus

Anatomy and Physiology

The zonular apparatus is a three-dimensional structure composed of 220-350 bundles of fibers that anchor and support the lens in the patellar fossa, facilitating accommodation by transmitting the contraction and relaxation of the ciliary muscle fibers. These fibers have a ring-like shape, with a triangular cross-section, and are synthesized by the non-pigmented ciliary epithelial cells in the pars plana.

The zonular apparatus is divided into four regions: pars orbicularis, zonular plexus, zonular bifurcation, and zonular limbs (anterior, equatorial, and posterior). The fibers are organized radially and circumferentially, most originating from the posterior part of the pars plana and merging with the basement membrane of the non-pigmented ciliary epithelium.

Zonular fibers are divided into three structural groups (21): anterior zonule (between pars plana and pre-equatorial area of the lens), posterior zonule (between pars plicata and post-equatorial area), and equatorial zonule (between pars plicata and lens equator). The insertion of zonular fibers on the lens occurs pre-equatorially, equatorially, and post-equatorially, each with distinct characteristics. Although anatomically zonular fibers can be classified separately, they form a continuous three-dimensional network essential for the efficient functioning of the lens and the accommodation process (22).

Pathophysiology

In Marfan syndrome, ocular disorders, especially ectopia lentis, are essential for diagnosis (23). Ectopia lentis represents the displacement of the lens from its normal anatomical position due to zonular abnormalities that can be genetic (Marfan syndrome, Weill-Marchesani syndrome) or acquired (trauma, uveitis).

Ectopia lentis causes symptoms (24) such as blurred vision, monocular diplopia, headache, and dizziness, and can lead to severe complications such as secondary glaucoma, phacolytic uveitis, and retinal detachment. In Marfan syndrome, lens subluxation is usually bilateral and symmetrical, occurring in the first 20 years of life and stabilizing thereafter (24).

Zonular impairment in Marfan syndrome is due to fibrillin-1 deficiencies, a major component of the microfibrils in ocular connective tissues. This leads to suboptimal zonular anchorage and lens dislocation (25). Histologically, zonular fibers show thinning, elongation, and irregular diameter (26), and ultrastructurally, a reduced number of fragmented and disorganized zonular fibers (27). In addition, ciliary processes are fewer (28) and zonules are susceptible to the proteolytic action of certain enzymes, contributing to lens instability (29).

1.3. Molecular pathogenesis of fibrillinopathy in Marfan syndrome

Connective tissue provides anchorage, mechanical strength, elasticity, and reversible deformability to various anatomical structures. The extracellular matrix, the main component of this tissue, includes elastic and collagen fibers in a hydrophilic gel formed by proteoglycans and glycosaminoglycans (30). Fibrillin is the main structural component of the microfibrils in the extracellular matrix (31), playing an essential role in the process of elastogenesis and maintaining the structural integrity of tissues.

Roles of fibrillin-1: formation of mature elastic fibers (32,33); connecting elastic fibers with other components of the extracellular matrix (34); providing elasticity and resistance of tissues to traction (35); tissue homeostasis through interaction with the latent TGF-ß complex and bone morphogenetic proteins (34,36).

Mutations in the FBN1 gene

Marfan syndrome is caused by mutations in the FBN1 gene, which encodes fibrillin-1 (37). Mutations affect the synthesis, processing, and assembly of fibrillin in microfibrils. Altered fibrillin-1 is not properly secreted, does not incorporate into the matrix, or is present in reduced amounts (38). There have been 3000 mutations of the FBN1 gene reported (39), most of which are unique to an individual or family (40).

Genotype-phenotype correlation is difficult due to the large number of mutations and clinical variability (41,42). Mutations in the exon 24-32 region are associated with the severe neonatal phenotype (43). Mutations in the exon 23-29 region are associated with Marfan syndrome without ocular involvement (44).

Clinical manifestations:

Marfan syndrome affects 1 in 5000 individuals (45). Manifestations include ocular, cardiovascular, musculoskeletal, integumentary, and pulmonary abnormalities. Ocular involvement is more common with mutations involving in-frame deletions and insertions or missense mutations with cysteine substitution (46). The integrity of the extracellular matrix and local TGF-ß activity are compromised, explaining the systemic manifestations of Marfan syndrome (35).

1.4. Ocular manifestations and diagnostic algorithm of Marfan syndrome

Common ocular manifestations:

Ectopia lentis: has a prevalence of 60% (2), often detectable from birth or in childhood (47). It manifests as blurred, fluctuating vision and monocular diplopia. Myopia: 34-44% of patients present with moderate to high myopia (48). It can be axial, with an anteroposterior axis longer than the population average (49).

Diagnostic algorithm of Marfan syndrome:

- Berlin Nosology (1986): Initial set of clinical criteria (50).
- Ghent Nosology (1996): Revision of the Berlin criteria to include family history, but found to favor false-positive diagnosis (50).
- Revised Ghent Nosology (2010): Includes clinical criteria, family history, and genetic testing, with standardized diagnostic rules and causality criteria for FBN1 mutations (23).

Depending on the presence or absence of family history, the diagnostic algorithm is as follows (23,51):

- without family history: enlarged aortic diameter/aortic dissection + ectopia lentis; enlarged aortic diameter/aortic dissection + FBN1 gene mutation; enlarged aortic diameter/aortic dissection + systemic score ≥ 7; ectopia lentis + disease-causing FBN1 mutation.

- with family history: ectopia lentis; systemic score ≥ 7; enlarged aortic diameter with Z-score ≥ 2 (over 20 years) and Z-score ≥ 3 (under 20 years).

Systemic score (23): Clinical manifestations receive a score (e.g., "thumb" sign - 3 points, myopia >3D - 1 point). A score ≥ 7 indicates systemic involvement.

Ocular criteria of the revised Ghent nosology (23): ectopia lentis and myopia are the most important ocular features; myopia > 3D contributes to the systemic score. Ectopia lentis must be carefully evaluated and repeated if lens subluxation is not evident.

Differential diagnosis (23) with conditions with ectopia lentis: familial ectopia lentis, Weill-Marchesani syndrome, homocystinuria, Stickler syndrome.

2. Current surgical management of lens subluxation (ectopia lentis) in marfan syndrome with and without the use of capsular tension rings

The surgical management of ectopia lentis has advanced significantly due to modern devices (sutures, capsular retractors, rings and capsular tension ring segments, and artificial lenses) that improve outcomes and postoperative prognosis. Lens subluxation presents a major surgical challenge due to instability, necessitating the use of temporary or permanent support devices.

2.1. Medical devices used in current surgical management of ectopia lentis

2.1.1. Capsular retractors

These manage zonular dehiscence intraoperatively. They prevent capsular bag aspiration, provide anterior-posterior support, and offer torsional stability (52).

2.1.2. Capsular tension rings (standard, cionni-type, segments, alternative devices)

Introduced in 1991 (53), capsular tension rings have evolved to maintain the contour of the capsular bag after lens removal. Modern models, made from PMMA, offer improved stability and adapt to different capsular bag sizes. There are various types of rings: standard (CTR), modified (MCTR), and segments (CTS), each having specific uses depending on the severity of the subluxation and patient needs.

The action principle of tension rings (54) involves distributing centrifugal force evenly along the circumference of the capsular bag, recentering the bag, and stabilizing the artificial lens. They are useful in cases of mild to moderate zonular dehiscence. Modified rings, such as the Cionni-type, are sutured to the sclera to provide additional stability in cases of severe zonulopathy.

Standard Capsular Tension Rings (CTR)

CTRs are open rings with an oval cross-section and loops at both free ends (54,55), a design that allows for safe insertion and placement of secondary devices (55). Indications for using CTR (56) include mild zonulysis (< 4 clock hours) and mild generalized zonular laxity. Implantation can be done manually with forceps or with a disposable or reusable injector (54).

The Henderson ring (57) is a specially designed CTR with eight equally spaced indentations that allow for the removal of nuclear and cortical material while maintaining tension on the equator of the capsular bag.

Selection of sizes is based on the dimensions of the capsular bag, correlated with the axial length of the eye and corneal diameter (58,59). A size is preferred that ensures overlap of the terminal loops for maximum circumferential support.

Advantages: it stabilizes the artificial lens by reducing centripetal contraction of the capsule and capsulorhexis (60).

Disadvantages: it does not provide anterior-posterior support for the capsular bag or rotational countertraction (61) and may hinder cortical aspiration by trapping the cortex in the equator of the capsular bag (57).

Contraindications (55): posterior capsule ruptures with vitreous loss, incomplete or unstable capsulorhexis, very small eyes with a very narrow iridocorneal angle (risk of angle closure).

Cionni Modified Capsular Tension rings (MCTR)

In cases of extensive zonulopathy and severe lens subluxation, standard capsular tension rings (CTR) are not sufficient to ensure adequate centering of the capsular bag. For these situations, Cionni designed in 1998 the modified capsular tension rings (MCTR), which allow for effective anchoring of the intact capsular bag to the scleral wall (62).

Characteristics and types of Cionni MCTR

MCTRs are made from PMMA and have an open-ring shape. Unlike standard CTRs, Cionni-type MCTR are equipped with one or two fixation loops on their circumference, through which the ring is sutured to the sclera (63–65). These loops protrude 0.25 mm and are positioned anterior to the anterior capsule, thus preserving the integrity of the capsular bag during suturing (54,63–65).

Models of Cionni MCTR (63–65):

- 1-L: Features the anchoring loop to the sclera located at the distal end of the ring.
- 1-C: Features the anchoring loop located at the proximal end of the ring.
- 2-L: Has two fixation loops, providing additional stability.

Advantages: it provides secure and stable anchoring of the capsular bag to the scleral

wall, being useful in cases of severe zonular insufficiency and preserving the integrity of the capsular bag due to the anterior positioning of the fixation loops (66).

Disadvantages: the complex design of the rings with two loops makes them difficult to implant into the capsular bag.

Requirements for effective fixation (54): to ensure effective fixation using MCTR, a properly sized capsulorhexis (5-6.5 mm) is necessary. This allows for safe cataract removal, stable positioning of the ring, and proper interference between the MCTR loop hook and the edge of the capsulorhexis.

Malyugin (67) developed a variant of the MCTR with a spiral-shaped end of the ring, allowing the ring to slide along the equator of the capsular bag, reducing the risk of bag perforation. The spiral shape allows intraocular insertion of the ring using an injector.

Capsular Tension Segments (CTS)

Designed by Ahmed in 2002 for patients with extensive and/or progressive zonular dehiscence (55). They are made from PMMA and have a 120° arc shape with radii of curvature of 4.5 mm (model 6E) or 5 mm (model 6D) and have a single fixation loop placed anteriorly (54). Segments with a 120° arc shape do not generate centrifugal force over 360°, characteristic of CTR and MCTR. They can be used in cases of discontinuous anterior capsulorhexis and posterior capsule ruptures. Multiple segments can be used to secure a larger circumference, providing support only in the transverse plane. For circumferential support, segments must be combined with CTR or MCTR (54). They are placed in the area of maximum zonular dehiscence and are sometimes used only as intraoperative support, being removed at the end of the procedure.

Alternative devices for stabilizing the capsular bag

Capsular anchor (Hanita Lenses) (68): made from PMMA, it has an anchor-like design allowing suturing to the sclera. Recommended for moderate and severe zonular dehiscence. Anchors the anterior capsule to the sclera.

Bean-shaped ring segment (69): made from PMMA, with an inner diameter radius of 2.5 mm and outer diameter radius varying between 5.5 mm and 7.0 mm. Used in association with bag-in-the-lens lenses, available in a simple variant and with a scleral anchoring loop.

Ambati capsular tension segments (70): unlike Ahmed's segments, these have two fixation loops to the scleral wall. They distribute tension at two points, reducing the risk of complications associated with transmitting tension at a single point, such as anterior capsule ruptures.

Capsular tension segments (CTS) and various alternative devices for stabilizing the

capsular bag provide specialized solutions for cases of extensive or severe zonular dehiscence. The choice of the appropriate device depends on the particularities of each case and the surgeon's preferences, considering the need to provide optimal support and minimize the risks associated with surgical intervention.

2.1.3. Sutures and artificial lenses

Sutures selection is based on criteria such as tensile strength, tissue response, ease of handling, absorbability, and size. For the cornea and sclera, which have no blood circulation, durable sutures that do not induce chronic inflammation are preferred (71).

Nylon 10-0, frequently used for corneal, scleral, and conjunctival sutures; loses strength after 12-18 months. Prolene 10-0, preferred for permanent sutures, such as suturing artificial lenses or the iris; disadvantages include difficulty in handling and tissue erosion over time.

To avoid issues related to suture degradation in permanent sutures, thicker polypropylene sutures (9.0 or 5.0) have been adopted for securing lenses and other medical devices (72).

Artificial lenses - cataract surgery until 1949 resulted in aphakia. Today, artificial lenses are classified by material, haptic appearance, optic, and intraocular positioning (73). In Marfan syndrome, anterior chamber artificial lenses were used for a long time, but later, lenses implanted in the capsular bag anchored to the sclera were preferred.

2.2. Surgical techniques with preservation of the capsular bag stabilized with a modified cionni-type capsular tension ring

Preserving the capsular bag and implanting the lens inside it are essential for maintaining the anatomical order of intraocular structures and represent the main goal in surgical interventions. Modern devices (capsule retractors, tension rings) and viscoelastic substances facilitate achieving this goal.

The Cionni technique, described by Cionni in 2003, involves the following steps (74): regional anesthesia followed by a primary incision in the clear cornea, in 3 planes. Two types of substances are injected, one to stabilize and push the vitreous and the other to maintain the depth of the anterior chamber. Capsulorhexis is performed under viscoelastic protection, followed by a 1 mm incision for introducing an iris retractor, stabilizing the lens for hydrodissection.

Phacoemulsification is done in the anterior chamber with low parameters, and the lens cortex is removed with the aid of viscodissection. A 9.0 Prolene suture is introduced through the loop of a 1-L tension ring, which is manipulated into the capsular bag to place the anchoring loop above the capsulorhexis margin. A scleral pocket is created, and the sutures

are introduced through the sulcus and sclera, and the thread is tied for centering. An artificial lens is implanted under viscoelastic protection. The substances are washed, the incisions are hydrated, and the conjunctiva is sutured.

Variants and innovations:

-anchoring MCTR with 5.0 polypropylene suture (72)

-use of capsular fixation segments as an alternative to tension rings, providing stability in cases of severe zonulopathy (75)

2.3. Techniques without preservation of the capsular bag

In cases of severe subluxation where the capsular bag cannot be preserved, the artificial lens is implanted using one of the following techniques:

- Anterior chamber implantation (76): the lens is placed in the anterior chamber after vitrectomy and pharmacological miosis.
- Iris-clipped artisan lens (77): the lens is fixed to the iris after removing the capsular bag and vitrectomy.
- Iris-sutured artificial lens (78): the procedure involves suturing the lens to the posterior surface of the iris.
- Sclera-sutured artificial lens (79): after peritomy and creating scleral flaps, the artificial lens is fixed with Prolene sutures to the sclera.

II. Personal contributions

3. Specific objectives and working hypothesis

Marfan syndrome affects 1 in 5000 individuals (45) and can cause various ocular problems, the most common being ectopia lentis, present in approximately 60% of cases (2). When lens subluxation can no longer be corrected with glasses or contact lenses, surgery for lens ectopia is recommended.

Study hypothesis: the use of modified cionni tension rings for stabilizing the capsular bag is a reproducible technique that provides stability and proper centration of the implanted lens, resulting in postoperative visual acuity superior to other techniques.

Specific objectives:

- Compare the surgical techniques used for ectopia lentis.
- Evaluate the benefits brought by modified Cionni tension rings.
- Compare surgical techniques in children versus adults.
- Identify the most frequently used technique and associated complications.
- Correlate surgical techniques with postoperative visual outcomes.

- Assess the influence of surgical technique on the type of implanted lens (monobloc/three-piece, toric/non-toric).

Aspects investigated in the patients included in the study:

- Gender distribution of patients undergoing surgery.
- Age of patients at the time of surgery and the influence of age on the choice of surgical technique.
- Pre- and postoperative visual acuity.
- Frequency of resolving lens subluxation with modified Cionni rings.
- Distribution of biometric data (axial length, keratometry) compared to the specialized literature.
- Presence and degree of preoperative corneal astigmatism.
- Analysis of cases requiring toric artificial lens implantation.
- Analysis of the patient cohort as a whole and by age and gender categories.

4. Patients and methods

4.1. Patient selection

The study included 19 patients (33 eyes) with Marfan syndrome and an indication for surgery for ectopia lentis from the personal cases of Prof. Dr. Călin Petru Tătaru. The retrospective study was conducted between February 2012 and June 2020 at the Emergency Clinical Hospital for Ophthalmology in Bucharest, in accordance with the provisions of the Declaration of Helsinki and approved by the hospital's Ethics Committee.

All patients provided written consent for evaluation. Patients with satisfactory visual acuity obtained through optical correction, those with insignificant subluxation, very young patients, and those with advanced glaucoma, ocular inflammation, pterygium, keratoconus, or retinal diseases were excluded.

4.2. Preoperative evaluation

All patients had a confirmed diagnosis of Marfan syndrome associated with ectopia lentis, made in collaboration with a pediatrician, cardiologist, and sometimes genetic testing. Diagnosis was easier in families with a history of Marfan syndrome.

For surgery, adult patients presented with a complete set of tests and a cardiology clearance. Pediatric patients required a pediatric clearance confirming clinical health and a complete set of tests. During hospitalization, pediatric patients were accompanied by a parent or legal representative.

Upon admission, all patients underwent the following investigations:

autokeratorefractometry, visual acuity measurement, intraocular pressure measurement, biomicroscopic examination, fundus examination, biometry, and sometimes ultrasonography.

Autokeratorefractometry is the method used for evaluating total refraction and anterior corneal astigmatism in cooperative adult patients. The instrument used for this evaluation is the autokeratorefractometer, such as the one from Topcon. The procedure begins with the patient seated, with their chin on the lower support and forehead on the upper support of the device, looking at a fixed point, such as a house or balloon. Measurements are automated and performed three times for each eye, with the option to select manual mode. In pediatric or less cooperative adult patients, a portable autorefractometer, such as the Retinomax, is used.

Visual acuity measurement is the ability to distinguish details, measured by identifying high-contrast letters or symbols (black on white). The Snellen system is the most common, based on the minimum angle of separation between distinct objects visible to an emmetropic eye without refractive errors (80). Testing is done with each eye separately, at different distances, with or without optical correction, using letters from standardized Snellen charts. In cases of extremely low vision, acuity is noted by the ability to perceive hand movement, light, or the number of fingers. Newer systems, such as the Minimum Angle of Resolution (MAR) and logMAR (logarithm base 10 of MAR), are used for more precise measurements, with charts like Bailey-Lovie or ETDRS, featuring letters that progressively decrease in size in logMAR units.

Intraocular pressure measurement - indentation or applanation tonometry is used. In the study, intraocular pressure was measured with the Maklakov tonometer. The procedure involves applying a weight to the cornea, which leaves an imprint on the ceramic side of the weight inversely proportional to the intraocular pressure. It is essential for diagnosing and monitoring glaucoma, especially in Marfan syndrome patients, who are predisposed to various ophthalmic conditions.

Biomicroscopic examination is essential in ophthalmology for diagnosing various ocular conditions. This procedure uses a biomicroscope, a complex instrument with a binocular microscope that allows magnified observation of the eye (5X-40X), adjustable for the examiner's needs. The biomicroscope has an adjustable illumination system and various filters for detailed examination of ocular structures. The eyelids are examined first, followed by the conjunctiva, sclera, cornea, anterior chamber, iris, and lens. Lens subluxation is better highlighted on a dilated pupil and sometimes only noticeable by asking the patient to look in primary positions.

Fundus examination is best performed on a dilated pupil, using a biomicroscope and special lenses or an ophthalmoscope. The optic nerve, macula, vascular arcades, and retinal periphery are inspected.

Biometry, in the study, used two methods: acoustic biometry (ultrasound) and optical biometry. In adult patients and cooperative children, optical biometry was performed, which provided additional data beyond axial length: keratometric values, anterior chamber depth, corneal diameter, and lens thickness. The IOLMaster was used in the study. For uncooperative patients, acoustic biometry was performed under sedation, with simultaneous keratometry measurement using a portable autokeratorefractometer.

Data on axial length, anterior chamber depth, and lens thickness were obtained. The Ultrascan Eyes Scanner was used in the study. Repeated measurements were performed in both categories of patients to minimize errors.

Ultrasound was used in cases of complete lens opacities or pupils that do not dilate pharmacologically, preventing fundus examination.

4.3. Anesthesia

In the study, adult patients were operated on under retrobulbar anesthesia (involving the injection of anesthetic into the periocular muscle cone), while pediatric patients underwent surgery under general anesthesia with orotracheal intubation.

4.4. Surgical techniques used in the study

The resolution of ectopia lentis in Marfan syndrome is a complex problem, which is why ophthalmologists are constantly developing new devices and surgical techniques (3). Various shortcomings of equipment and the need for technically more accessible methods have led to experimenting with several types of surgical interventions.

Surgical techniques analyzed:

1. Implantation of a foldable artificial lens in the capsular bag with a standard tension ring.
2. Implantation of a foldable artificial lens in the capsular bag with a modified Cionni tension ring, anchored to the sclera at one point.
3. Implantation of a foldable artificial lens anchored to the sclera by a two-point suture.
4. Implantation of a foldable artificial lens with one haptic in the bag and one haptic anchored to the sclera by suture.

4.4.1. Implantation of a foldable artificial lens in the capsular bag with a standard tension ring

Instruments and consumables used: blepharostat, incision knives (1.2mm and 2.2mm), anterior chamber maintainer, various cannulas (for hydrodissection, aspiration,

hydrosuture), micro-scissors and forceps for capsulorhexis, viscoelastic substances, spatula, nucleus manipulator, tension ring, glass syringe, foldable artificial lens, injector, operating microscope, phacoemulsification apparatus, solutions and medicinal substances (gentamicin, adrenaline, Tobradex).

Surgical technique: after local or general anesthesia, the area is prepared with povidone-iodine solution, and a sterile field is applied. The blepharostat is placed to open the palpebral fissure (Figure 4.1.A). Corneal incisions are made at 5 o'clock (for the anterior chamber maintainer) and 2 o'clock (secondary incision), and a main incision at 9 o'clock. The anterior capsule is perforated with a disc or 23-gauge needle (Figure 4.1.B), followed by capsulorhexis with capsulorhexis forceps (Figure 4.1.C). The lens material is aspirated with the aspiration cannula or phacoemulsification apparatus (Figure 4.1.D), stabilizing the capsular bag with retractors if necessary. Under the protection of viscoelastic substances, the tension ring is inserted to stabilize the capsular bag (Figure 4.1.E). Subsequently, the artificial lens is folded and inserted into the capsular bag with the injector and Y manipulator (Figure 4.1.F). Finally, the viscoelastic substances are washed away, the incisions are closed with hydrosuture, antibiotic ointment is applied, and the patient is monitored postoperatively.

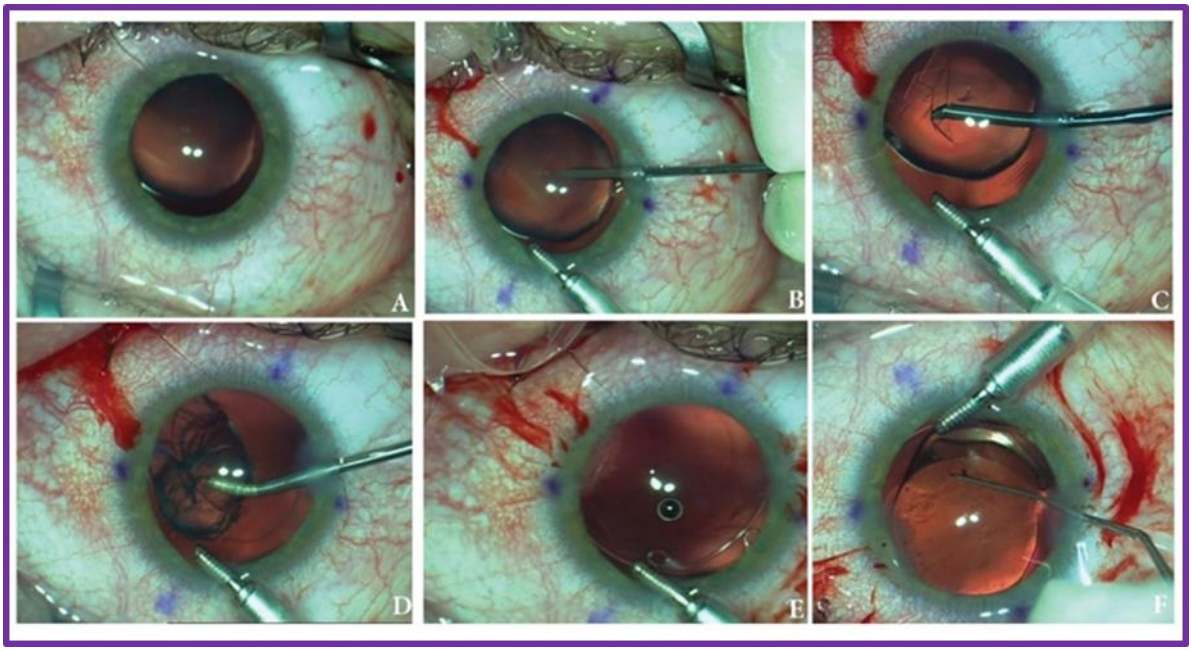

Figure 4.1. *Main operative steps of the technique involving artificial lens implantation in the capsular bag with a tension ring:* ***A****: Placement of blepharostat.* ***B****: Incision of the anterior capsule with a disc.* ***C****: Capsulorhexis with capsulorhexis forceps.* ***D****: Removal of lens material with an aspiration cannula.* ***E****: Insertion of a tension ring into the capsular bag.* ***F****:*

Implantation of an artificial lens in the capsular bag.

4.4.2. Implantation of a foldable artificial lens in the capsular bag with a modified cionni tension ring anchored to the sclera at one point

Instruments and consumables used: blepharostat, incision knives (1.2mm and 2.2mm), anterior chamber maintainer, various cannulas (for hydrodissection, aspiration, hydrosuture), micro scissors and forceps for capsulorhexis, viscoelastic substances, spatula, nucleus manipulator, tension ring, glass syringe, foldable artificial lens, injector, operating microscope, phacoemulsification apparatus, solutions and medicinal substances (gentamicin, adrenaline, Tobradex).

Surgical technique: Anesthesia, asepsis, and ocular antisepsis are performed. The patient is positioned correctly with the iris parallel to the floor (Figure 4.2.A). Using a 1.2mm knife, incisions are made for the anterior chamber maintainer, the secondary incision, and the main incision. The edges of the anterior capsule are perforated and reflected, creating a continuous capsulorhexis with forceps. After hydrodissection, the lens material is removed by aspiration or phacoemulsification (Figure 4.2.B). A scleral limbal incision is made, and a scleral pocket is created for anchoring the ring (Figure 4.2.C). The suture thread is passed through the scleral pocket into the eye and then externalized. The thread is passed through the anchoring loop of the ring. The Cionni ring is inserted intraocularly, and then the thread is passed back, being externalized 1.5mm from the first thread. The artificial lens is inserted into the capsular bag using an injector (Figure 4.2.D). The two threads are cut and knotted to center the lens (Figure 4.2.E).The incisions are hydrated and sealed (Figure 4.2.E), and the eye is covered with a compressive dressing.

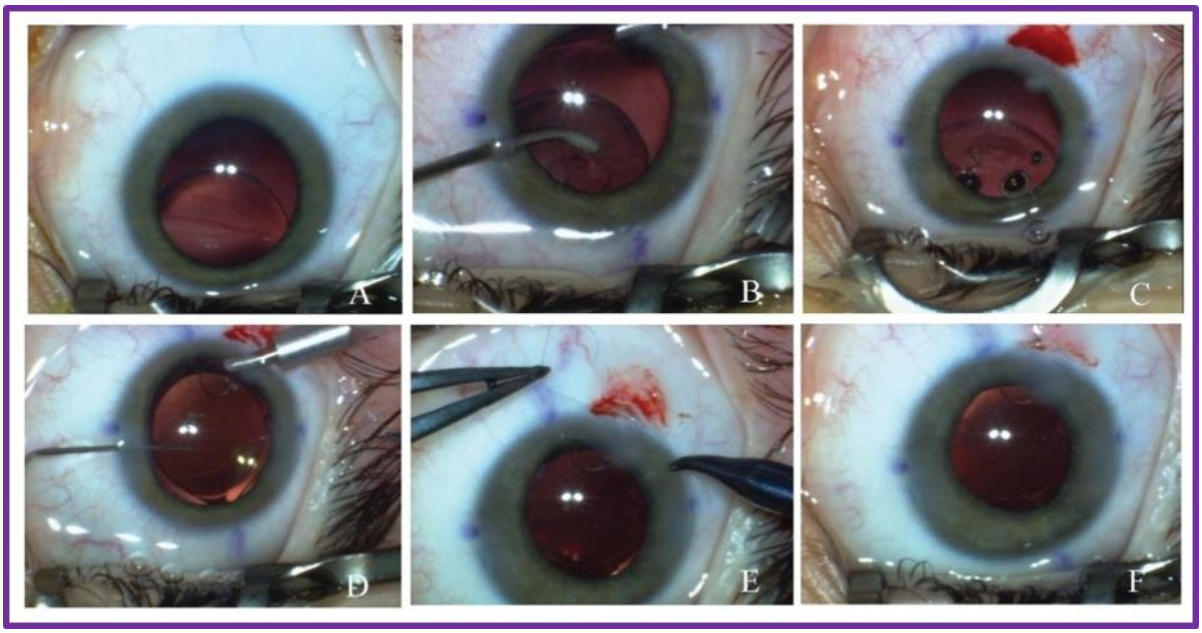

Figure 4.2. *Main operative steps of the technique involving artificial lens implantation with a modified Cionni tension ring anchored to the sclera at one point.*

A: *Initial image;* ***B:*** *Aspiration of the lens material;* ***C:*** *Creation of the scleral pocket;* ***D:*** *Implantation of the ring and lens in the bag;* ***E:*** *Knotting of the suture thread with centering of the bag-ring-artificial lens complex;* ***F:*** *Image at the end of the surgery*.

4.4.3. Implantation of a foldable artificial lens anchored to the sclera with a two-point suture

Instruments and consumables used: blepharostat; surgical knives of various sizes (1.2mm, 2.2mm); anterior chamber maintainer; various cannulas (for hydrodissection, aspiration, hydrosuture); forceps for capsulorhexis and other types of forceps; viscoelastic substances; manipulators and nucleus hooks; scissors and surgical scissors; suture threads and various types of needles; syringes and solutions for ocular irrigation; foldable artificial lens; operating microscope and phacoemulsification apparatus.

Surgical technique:

- Anesthesia, asepsis, and ocular antisepsis are performed, and a sterile field is placed (Figure 4.3.A).
- Two diametrically opposed points are marked on the sclera, 2.5mm from the limbus (Figure 4.3.B). A 1.2mm incision is made at the limbus for introducing the anterior chamber maintainer. Two more incisions are made, one for additional access and one enlarged to 2.2mm.
- The anterior capsule of the lens is perforated, and capsulorhexis is performed with forceps (Figure 4.3.C). After hydrodissection, the lens material is aspirated or emulsified using the phacoemulsification apparatus.
- The conjunctiva is dissected at the initially marked points for placing the suture threads. The polypropylene thread is passed through the sclera and tied to the haptics of the artificial lens (Figure 4.3.D).
- The artificial lens is implanted intraocularly and centered by tensioning the suture threads (Figure 4.3.E).
- The polypropylene threads are sutured to the sclera (Figure 4.3.F).
- The viscoelastic substances are washed from the anterior chamber, the incisions are hydrated and sealed (Figure 4.3.G).
- Drops and antibiotic and anti-inflammatory ointment are applied, and the eye is dressed.

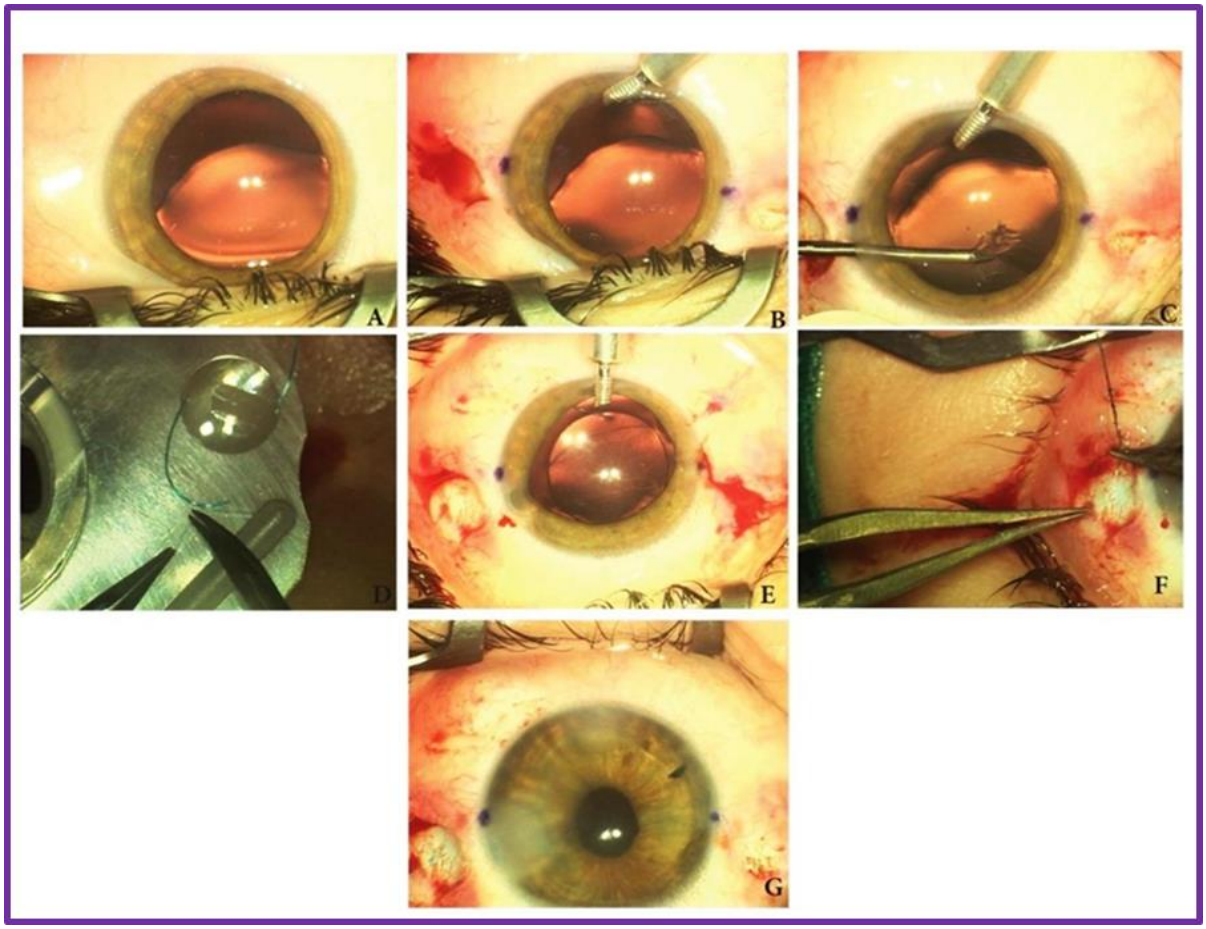

Figure 4.3. *Main operative steps of the operation involving the implantation of a foldable artificial lens anchored to the sclera with a two-point suture:* ***A:*** *Initial image;* ***B:*** *Two diametrically opposed points are marked on the horizontal axis;* ***C:*** *Performing capsulorhexis;* ***D:*** *Knotting the polypropylene thread on the haptic;* ***E:*** *Implantation of the artificial lens;* ***F:*** *Suturing the threads to the sclera;* ***G:*** *Image at the end of the surgery, before placing the threads on the conjunctiva.*

4.4.4. Implantation of a foldable artificial lens with one haptic in the bag and one haptic anchored to the sclera by suture

Instruments and consumables used: blepharostat, knives (1.2mm and 2.2mm), anterior chamber maintainer, disc knife, cannulas (for hydrodissection, aspiration, and hydrosuture), micro scissors and forceps for capsulorhexis, angled spatula, nucleus manipulator, "Y" hook, forceps, needle holder, small scissors, suture thread for lens and conjunctiva, viscoelastic substances, glass syringe, lens injector, operating microscope, phacoemulsification apparatus, syringes, saline solution, gentamicin, adrenaline, Tobradex ointment, infusion set, marker.

Surgical technique: After anesthesia and patient positioning, locoregional asepsis and antisepsis are performed with povidone-iodine, followed by preparation of the operative field. The area of maximum subluxation is marked, and then the incisions are made. After placing the maintainer, the anterior capsule is punctured, and capsulorhexis is continued with forceps. Hydrodissection of the lens is performed with a hydrodissection cannula and nucleus manipulation. The material is removed by manual aspiration or phacoemulsification. Conjunctiva dissection and suture for the first haptic of the artificial lens are performed. The lens is inserted into the cartridge and implanted intraocularly.The lens is positioned with one haptic sutured to the sclera and one haptic in the bag.The viscoelastic substances are aspirated, and the incisions are hydrated. The conjunctiva is sutured, and the ocular tone is checked. Tobradex ointment and a compressive dressing are applied.

4.5. Methods of statistical analysis

Statistical data processing in this retrospective observational analytical study was performed using the IBM SPSS v. 26.0 program (IBM SPSS, Armonk, NY, USA: IBM Corp).

The following parametric tests were used: t-test (Student's t-test), analysis of variance (ANOVA).

The non-parametric tests used were: Mann-Whitney U test, Wilcoxon Signed-Rank test, Kruskal-Wallis test.

The obtained data were compared with the control (*$p < 0.05$, **$p < 0.01$, ***$p < 0.001$).

5. Results

The results of the study involved the analysis of 33 eyes from 19 patients with Marfan syndrome and ectopia lentis, using IBM SPSS (Statistical Package for the Social Sciences) v. 26.0 (IBM SPSS, Armonk, NY, USA: IBM Corp). Of these 33 eyes, 28 were operated on bilaterally (14 patients) and 5 unilaterally (5 patients). The gender distribution was balanced, with 17 eyes from males and 16 from females. The average age at the time of intervention was 17.39 years, ranging from 4 to 42 years.

Out of the 33 eyes, biometric data (axial length and keratometry) was unavailable for one eye, possibly due to handling errors in the observation files in the archive. The average keratometric values for the entire group were: K1 of 40.72 D ± 2.37 D, K2 of 42.86 D ± 2.77 D, and Kmed of 41.83 D ± 2.46 D. Analyzing keratometric values by age groups, we

observed statistically significant differences in K1, K2, and Kmed ($p<0.05$). The two age groups were also analyzed based on the type of astigmatism (with-the-rule, against-the-rule, oblique), resulting in similar values, with with-the-rule astigmatism present in almost 80% of each group.

The anterior-posterior axial length of the eyeball was studied comparatively for both sexes and the two age groups, children/adults. For females, the average axial length was 23.46mm ± 1.05, with a minimum axial length of 21.20mm and a maximum axial length of 24.80mm. For males, the average axial length was 24.1mm ± 2.06, with a minimum axial length of 20.96mm and a maximum axial length of 27.30mm. The confidence interval was 95%, with a p-value threshold of 0.05.

Descriptive statistical analysis using the Wilcoxon test for axial length between the two sexes showed no statistical differences ($p=0.440$). For the analysis of axial length across the entire group and the two age groups, the axial length was divided into three categories: <22mm, 22-25mm, and >25mm.

In the first category, with an axial length of <22mm, 20% of children and 11.8% of adults were included, representing 15.6% of the total eyes. In the second category, with an axial length between 22 and 25mm, 66.7% of children and 76.5% of adults were included, with a percentage of 71.9% for the entire group. In the last category, with an axial length >25mm, 13.3% of children and 11.8% of adults were included, with an average for the entire group of 12.5%.

The values of the anterior-posterior axial length by age categories were analyzed using the Mann-Whitney U test, and there were no statistically significant differences ($p=0.213$).

From the descriptive analysis of the frequency of each technique across the entire studied group, it was found that the most frequently used technique was the implantation of an artificial lens in a bag stabilized with the help of a Cionni-type tension ring sutured to the sclera (66.7%).

The second most frequently used technique was the implantation of an artificial lens sutured to the sclera at two points (15.2%);

A similar frequency (12.1%) was noted for the technique involving the implantation of an artificial lens inside a bag stabilized with a simple capsular tension ring.

The least used technique (6.1%) was the implantation of a lens with one haptic in the bag and one haptic sutured to the sclera (Figure 5.1).

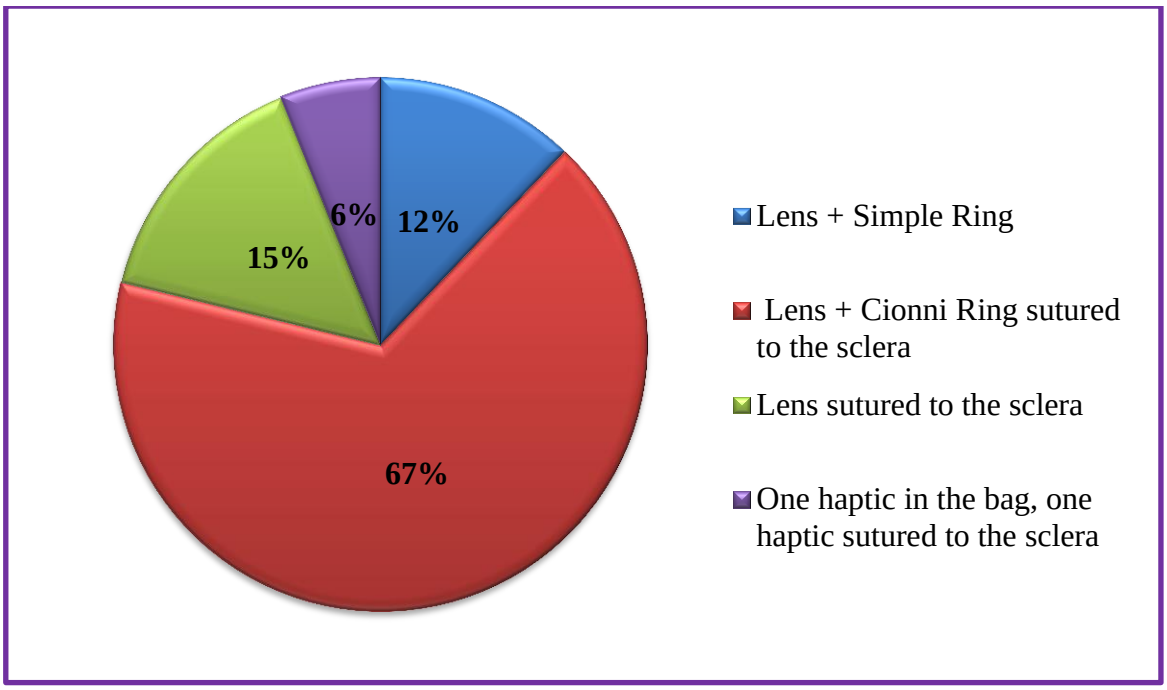

Figure 5.1. Distribution of Surgical Techniques Used in the Studied Cohort

The technique involving the implantation of an artificial lens together with a modified Cionni tension ring was the most frequently used in both children and adults (Table 5.1). Each operated eye was categorized into one of the following groups:

- **Group 1** – Technique involving the implantation of an artificial lens together with a simple capsular tension ring.
- **Group 2** – Technique involving the implantation of an artificial lens together with a Cionni-type capsular tension ring sutured to the sclera.
- **Group 3** – Technique involving the scleral suturing of an artificial lens.
- **Group 4** – Technique involving the implantation of an artificial lens with one haptic in the bag and one haptic sutured to the sclera.

Table 5.1. Frequency of each technique used in children and adults.

Statistical analysis of the techniques used in the study				
	Group 1 (n:4)	**Group 2 (n:22)**	**Group 3 (n:5)**	**Group 4 (n:2)**
Adults (n:17)	**4**	**11**	**1**	**1**
Children (n:16)	**0**	**11**	**4**	**1**

The distribution of the type of artificial lens used (monoblock/3-piece, toric/non-toric) for each group of surgical techniques, as well as the type of toric/non-toric lens implanted according to the value of astigmatism, is as follows:

The design of the implanted lenses was distributed as follows:

- 1-piece lens in 26 cases, of which 4 were in group 1 and 22 in group 2 of techniques, and none in groups 3 and 4;
- 3-piece lens in 7 cases: 5 from group 3 and 2 from group 4 (Figure 5.2).

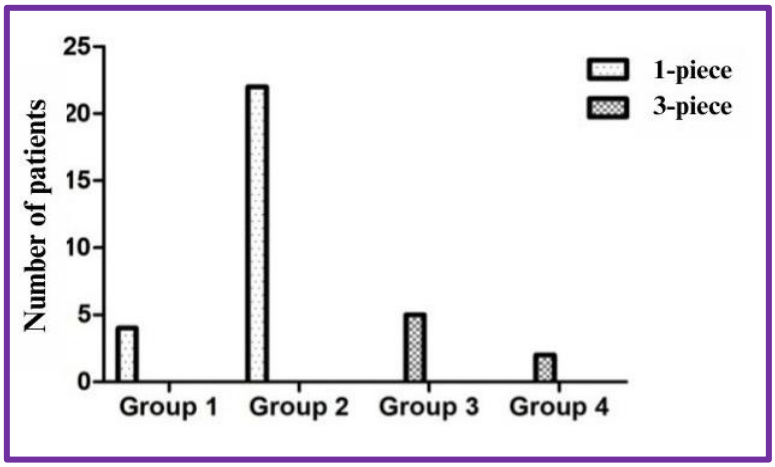

Figure 5.2. Distribution of 1-piece/3-piece lens type for each technique group.

Analyzing the correction of astigmatism for each surgical technique group revealed that:

- Toric lenses were implanted in 4 eyes from group 1, in 3 eyes from group 2, and in none from groups 3 and 4.
- Non-toric lenses were implanted in 19 eyes from group 2, in 5 eyes from group 3, and in 2 eyes from group 4 (Figure 5.3).

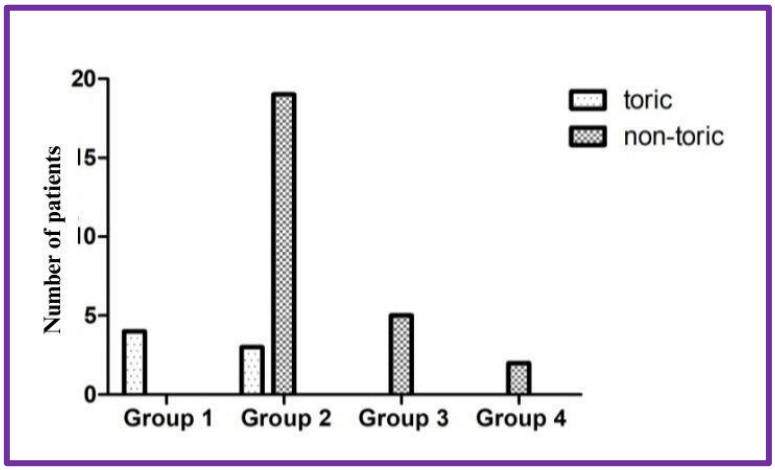

Figure 5.3. Distribution of the type of toric/non-toric lens in each technique group.

Preoperative astigmatism correction using toric artificial lenses was possible in 6

cases, all with cylinder values >2.50 (Figure 5.4).

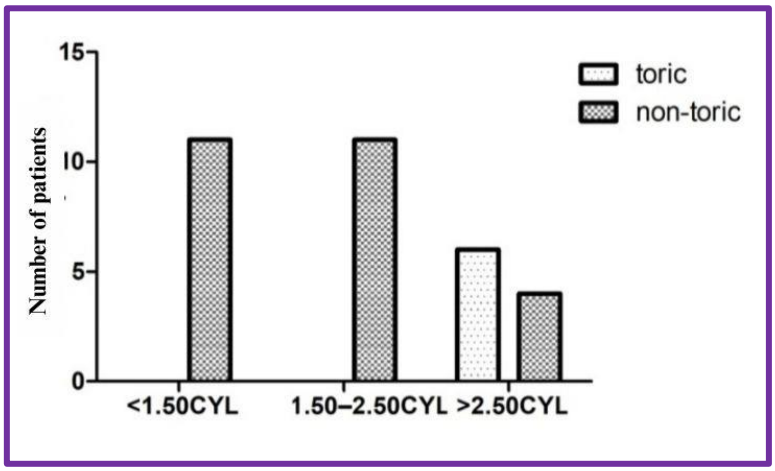

Figure 5.4. Type of toric/non-toric lens implanted based on cylinder value.

Preoperative visual acuity, measured in decimals (Snellen), was evaluated for all subjects. Postoperative visual acuity was assessed in 31 subjects, with 2 subjects lost to follow-up. Using the Wilcoxon test and ANOVA, we compared preoperative visual acuity with postoperative results for the entire group and found highly statistically significant differences with a p-value of 0.001. This indicates that visual acuity significantly improved postoperatively, despite missing postoperative data for 2 patients (Figure 5.5).

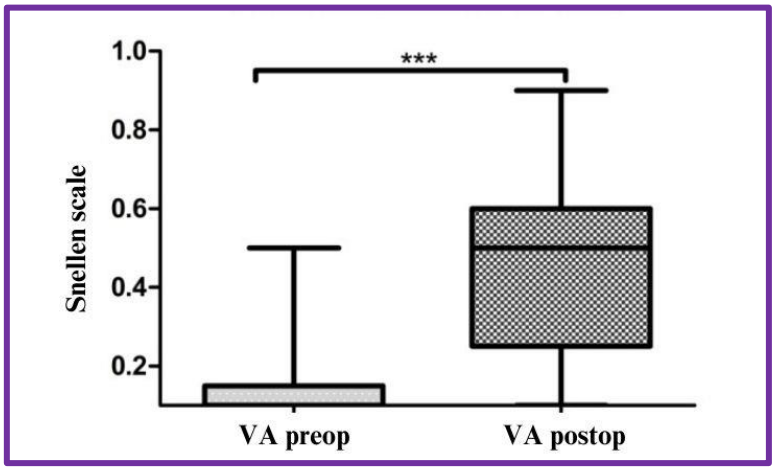

Figure 5.5. Visual acuity progression for the entire group.

We also analyzed the frequency of each postoperative visual acuity for each surgical technique used.

The minimum visual acuity of 0.1 was achieved in two subjects who underwent the technique of implanting a lens sutured to the sclera at two points. The maximum visual acuity

of 0.9 was also achieved in a subject operated on using the technique of implanting a lens sutured to the sclera at two points. Following the surgery, out of the 31 available visual acuities, 18 eyes achieved a visual acuity ≥0.3 but <0.8. Four eyes had a visual acuity ≥0.8. Eyes from the group 2 technique presented the most normal or nearly normal visual acuities (a total of 14 eyes).

Due to the unequal distribution of the number of patients operated on with each technique, with a significantly higher number in group 2, we statistically analyzed this group against the other 3 groups combined (Table 5.2). In this comparison, visual acuities were expressed in logMAR.

Table 5.2. Statistical analysis between techniques with mean descriptive statistics (Mann-Whitney, Kruskal-Wallis)***

Statistical analysis between technique 2 and the average of techniques 1, 3, and 4			
	Technique 2 (22 eyes)	**Avergae of technique 1,3, 4 (11 eyes)**	***p* ***
Age	**17.90 ±12.69**	**16.36 ± 10.08**	**0.818**
AL	**23.55 ± 1.06**	**24.26 ± 2.40**	**0.751**
BCVA preop	**0.91 ± 0.25**	**0.94 ± 0.31**	**0.643**
BCVA postop	**0.41 ± 0.22 ** 0.0001**	**0.41 ± 0.33 **0.005**	**0.798**
IOL	**1–piece in all cases**	**3–piece in 7 cases and 1-piece in 4 cases**	
Complications	**1 major**	**1 minor**	
IOP	**17.31 ±1.32**	**17.7± 4.13**	**0.567**
K mean	**41.96 ± 2.20**	**41.59 ± 3.01**	**0.678**

No differences were observed in terms of age, astigmatism, or intraocular pressure between group 2 and the other 3 groups. The type of lens implanted (monoblock or 3-piece) was implanted in all cases from group 2 and only in 4 cases from the other groups. The table also shows that there are no statistically significant differences regarding age between the two groups (p = 0.818, Kruskal-Wallis test*), axial length (p = 0.751, Kruskal-Wallis test*), or preoperative visual acuity (p = 0.643, Kruskal-Wallis test*), and no statistically significant differences in postoperative visual acuity between the two groups (p = 0.798, Kruskal-Wallis test*). However, statistically significant differences were found for each technique in terms of preoperative and postoperative visual acuity.

6. Discussions

Marfan syndrome, with an incidence of 1:5000 individuals (45), is associated with lens subluxation (ectopia lentis), which can significantly reduce visual acuity and poses a major therapeutic challenge. Surgical intervention is primarily indicated for decreased visual acuity, but it is difficult to assess in young children and impossible in non-verbal ones. Therefore, sometimes a passive approach is chosen over radical surgical intervention.

In the studied group, the average age at the time of surgery was 17.39 years, with a minimum age of 4 years and a maximum age of 42 years (81). Dividing the group by age into two categories, adults and children, with an almost equal distribution of subjects in the two categories, allows us to conclude that we have a homogeneous group in this respect.

In choosing the surgical technique, several aspects were considered: the patient's age, the degree of ectopia lentis, and the surgeon's experience and preferences. For all operated eyes, the preoperative technical objectives were to preserve the anatomical barrier, represented by the capsular bag, and to implant the artificial lens in the capsular bag.

Preserving the capsular bag and maintaining the normal anatomy of the eye is considered beneficial for patients by reducing the rate of complications (retinal detachment, glaucoma, vitreous loss, decentration of the artificial lens (82)).

Thus, out of a total of 33 eyes, successful implantation of the artificial lens in the sac and centering of the capsular bag-lens complex using Cionni-type capsular tension rings sutured to the sclera was achieved in 22 eyes (81). This technique can be associated with the late occurrence of breaking or biodegradation of the polypropylene suture anchoring the ring to the sclera.

Vote presented a study on 61 eyes where posterior chamber lenses were sutured to the

sclera using 10.0 polypropylene sutures (83). The most common late complication was suture breakage, occurring in 26.2% of eyes. Other authors contradict Vote's conclusions and results, considering polypropylene sutures stable and safe, with a breakage risk of less than 0.5% (84).

Postoperative visual acuity for each technique shows that out of the 31 visual acuities evaluated, 22 eyes had normal or near-normal visual acuities, i.e., postoperative VA ≥ 0.3, a value adopted at the International Congress of Ophthalmology in 2002 (85).

If the eyes from group 2 techniques had the most normal or near-normal visual acuities (a total of 14 eyes), the best visual acuity of 0.9 and the worst visual acuity of 0.1, considered severe vision loss (85), were observed in eyes from group 3 techniques. These data coincide with Maharana's conclusions (86) that the degree of lens subluxation at the time of surgery does not influence the postoperative visual outcome.

A common feature of patients with Marfan syndrome is an increased anteroposterior axis (87). Subjects in the doctoral study also had a longer average anteroposterior axis for each sex (24.13 mm in males and 23.46 mm in females) than those without Marfan syndrome (23.64 mm in males and 23.23 mm in females (88)). Comparing with Olsen (89), no statistically significant difference was found in the anteroposterior axis lengths between males and females in the study group.

The keratometric measurements of eyes included in the doctoral study are similar (81) to those obtained by other authors in the literature. Thus, the average Kmed values were similar to those presented by Gehle (49) in his extensive work involving 285 patients with Marfan syndrome.

The preoperative astigmatism value was obtained by subtracting the K1 value, expressed in diopters, from the K2 value, also expressed in diopters. The presence of average cylinders > 1.5D in both sexes confirms Konradsen's observations (90) that patients with Marfan syndrome associated with ectopia lentis present greater astigmatism compared to those without ectopia lentis.

Preoperative astigmatism correction through the implantation of toric artificial lenses improves postoperative visual acuity. In the studied group, only non-toric artificial lenses were implanted for cylinder values ≤ 2.50D Cyl, while toric artificial lenses were successfully implanted in 6 cases for >2.50D Cyl, and in 4 cases, non-toric artificial lenses were implanted (81).

Toric artificial lenses were implanted only in cases where the sac was preserved and stabilized with a simple tension ring or a Cionni-type ring anchored to the sclera (81).

The artificial lenses used were classified based on design into monoblock (1-piece) and 3-piece. The monoblock design was preferred in techniques where the crystalline sac was preserved and stabilized using simple rings or Cionni-type capsular tension rings sutured to the sclera (group 1 or group 2 techniques). This artificial lens design is specifically designed to be implanted in the capsular bag (91). Implantation in the sulcus can lead to pigment dispersion syndrome, glaucoma, recurrent ocular inflammation, macular edema, or vitreous hemorrhage (91). For the other 2 techniques (group 3 and group 4 techniques), only 3-piece artificial lenses were used, specially designed for sulcus placement (92).

During the 6-month follow-up of patients, 2 complications were encountered, one major, in an eye operated on with technique 2 (artificial lens implant with a Cionni-type capsular tension ring sutured to the sclera), and one minor, in an eye operated on with technique 1 (implantation of a lens in a sac stabilized with a simple ring was used). In the first case, the 10.0 polypropylene suture came undone and required another surgical intervention for repositioning. In the second case, the lens was slightly decentered, with no need for further surgical intervention. Fine opacifications of the posterior capsule were not noted as complications. It is possible that a longer follow-up period would increase the number of complications and reinterventions.

7. Conclusions and original contributions

Marfan syndrome is a rare but debilitating condition that primarily affects visual acuity due to ectopia lentis. Surgical correction of this condition is technically challenging, and there is currently no gold standard for treatment. The literature describes numerous surgical techniques, with or without preservation of the capsular bag and the implantation of an artificial lens. However, this study uniquely presents a comparative analysis of four different surgical techniques, with and without preservation of the lens capsule.

Retaining the lens capsule and strengthening the zonular belt with a Cionni ring anchored to the sclera has proven to be a very effective and reproducible technique. This technique was associated with only one major postoperative complication and improved visual acuity in every case.

Although scleral anchoring of the Cionni rings can be associated with the long-term rupture of 10.0 polypropylene sutures, this complication was not encountered in our study, possibly due to the short follow-up period. In the future, we might try anchoring the Cionni rings with thicker polypropylene sutures, such as 9.0.

While we cannot say there is a correlation between the surgical technique used and

postoperative visual acuity, the eyes in group 2 showed the most normal or near-normal visual acuities. Both the lowest visual acuity (0.1 Snellen) and the highest (0.9 Snellen) were obtained in eyes from group 3 techniques.

Synthesis of Personal Contributions

- Comparative evaluation: this study provides a comparative evaluation of four different surgical techniques used to treat lens subluxation in patients with Marfan syndrome.
- The technique involving the implantation of an artificial lens together with a Cionni-type capsular tension ring sutured to the sclera was most often feasible (22 out of 33 eyes) and proved to be an effective and reproducible method.
- Visual acuity improved in all patients, regardless of age or type of intervention. However, patients operated with the implantation of an artificial lens and a Cionni-type capsular tension ring sutured to the sclera achieved the most normal or near-normal visual acuities.
- The surgical techniques used in children versus adults are different due to specific factors affecting the timing of surgery. Adults showed a lower degree of lens subluxation, resulting in good visual acuity until older ages, making surgical techniques simpler (most often involving artificial lens implantation with a Cionni-type capsular tension ring sutured to the sclera or with a simple capsular tension ring).
- In children, significant subluxation accelerates the timing of intervention, and more frequently than in adults, it is necessary to suture an artificial lens to the sclera at two points.
- The surgical technique influences the design choice of the artificial lens, whether monobloc or three-piece.
- The surgical technique also influences the choice between toric and non-toric lenses.
- Complication rate of the 33 eyes operated on, there was only one major complication during the follow-up period, making it statistically insignificant.
- The timing of intervention for ectopia lentis is not standardized, with unsatisfactory visual acuity being the primary indicator for surgery. Given the improvement in visual acuity for all patients, we can conclude that the timing of the operations was optimally chosen.
- The study group was homogenous, with approximately equal numbers of patients by gender and age, ensuring the validity of the biometric data results.
- Keratometric values revealed the presence of flatter corneas in the study group,

confirming literature reports of flatter corneas in patients with Marfan syndrome.

- Astigmatism analysis indicated that with-the-rule astigmatism was most common.
- Astigmatism value influences the choice of toric or non-toric lenses.
- The mean axial length for the study group revealed a longer anteroposterior axis, confirming literature data. Axial length did not correlate with gender or age.
- Improvement of techniques for implanting capsular tension rings (96) .
- Innovative application for stabilizing toric lenses with Cionni-type capsular tension rings (97).

Limitations

- The relatively small number of cases was due to the incidence of Marfan syndrome in the population.
- There was no control group for parameter comparison.
- There was an unequal distribution of eyes operated on by each technique.
- The follow-up period for patients was short.
- Some parameters were missing due to their absence in the surgical records (possibly lost during file handling in the archive).
- The labor-intensive surgical techniques require complex and costly instrumentation.
- The surgeon's experience is an important factor in the success of each intervention, particularly in delicate cases.

Future directions

Starting from the working hypothesis for this study, which suggests that technique 2 (implantation of an artificial lens in a capsular bag stabilized with a Cionni-type capsular tension ring) is superior to the other techniques in terms of visual results, reproducibility, and long-term stability, the patient group should be expanded to ensure equal distribution of eyes operated on by each technique.

It would also be useful to evaluate this technique in comparison with other lens subluxation correction techniques described later. Among the recently described techniques applicable to ectopia lentis in Marfan syndrome, Canabrava's (72), Rossi's (93), and Yamane's (94) techniques are noteworthy.

Another development direction for the study could be the opportunity to replace the biodegradable sutures of the Cionni rings, whose resorption over time can lead to decentration of the artificial lens, with non-biodegradable sutures (such as PTFE), which would prevent this decentration (95).

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List of published scientific works

• First author/principal author SCIE – Web of Science (ISI) indexed journals:

1**. Dogaroiu AC**, Dudau M, Dogaroiu C, Tataru CP. Visual Outcomes in Ectopia Lentis in Marfan Syndrome: A Study of Four Surgical Techniques in Children and Adults. Medicina.

2024 Jul 5;60(7):1098. https://doi.org/10.3390/medicina60071098, IF 2.4
II. Personal contributions. Chapter 3. Specific objectives and hypothesis pp. 24-25 Chapter 4. Patients and methods pp. 26-73 Chapter 6. Discussions pp. 96-103 Chapter 7. Conclusions and personal contributions pp. 104-107
2. Tataru CP, **Dogaroiu AC***, Tataru CI, Dogaroiu C. Enhancing rotational stability of toric intraocular lenses using a type 2L Cionni capsular tension ring in patients with high myopia. J Cataract Refract Surg. 2019;45(9):1219–21. https://doi.org/10.1016/j.jcrs.2019.05.045, IF 2.6
II. Personal contributions. Chapter 4. Patients and methods pp. 26-73 Chapter 7. Conclusions and personal contributions pp. 104-107
3. Tataru CP**, Dogaroiu AC***, Mihai C. Modified injector for optimal insertion of standard CTRs in lax zonules. Eur J Ophthalmol. 2015 Jan;26(1):98–100. https://doi.org/10.5301/ejo.5000613, IF 1.4
II. Personal contributions. Chapter 4. Patients and methods pp. 26-73 Chapter 7. Conclusions and personal contributions pp. 104-107

- Coauthor SCIE – Web of Science (ISI) indexed journals:

1. Ní Dhubhghaill S, **Dogaroiu AC**, Zakaria N, Tassignon M-J. Modified bean-shaped ring segments for suture fixation of the bag-in-the-lens intraocular implant. J Cataract Refract Surg. 2017;43(8):1003–6. https://doi.org/0.1016/j.jcrs.2017.04.042, IF 2.6
I. General Part. Chapter 2. Current surgical management of lens subluxation (ectopia lentis) in marfan syndrome with and without the use of capsular tension rings pp. 14-23

- First author PubMed (BDI) indexed journal:

1. **Dogaroiu AC**, Dogaroiu C, Tataru CP. Ectopia lentis surgery in Marfan's syndrome: from "couching" to the use of intracapsular tension rings. Rom J Ophthalmol. 2022 May 2;66(1):2–7. https://doi.org/10.22336/rjo.2022.2
Introduction pp. 1-2; II. Personal contributions. Chapter 3. Specific objectives and hypothesis pp. 24-25; Chapter 4. Patients and methods pp. 26-73.

- First author chapter in international published textbook:

1. **Dogaroiu AC**, Ní Dhubhghaill S. Scleral anchoring of the modified bean-shaped ring segments. In Tassignon M-J, Ní Dhubhghaill S, Van Os L, editors. Innovative implantation technique. Bag-in-the-lens cataract surgery. Cham, Switzerland: Springer Nature;2019.p. 167-172.
I. General Part. Chapter 2. Current surgical management of lens subluxation (ectopia lentis) in marfan syndrome with and without the use of capsular tension rings pp. 14-23