Congenital Cataract Surgery: A Case Series from a Tertiary Center

Cirurgia de Catarata Congénita: Uma Série de Casos de um Hospital Terciário

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Recebido/Received: 2024-10-14 | **Aceite/Accepted:** 2025-07-28 | **Published online/Publicado online:** 2025-11-26 | **Publicado/Published:** 2026-03-25

DOI: https://doi.org/10.48560/rspo.38215

ABSTRACT

INTRODUCTION: Congenital cataract is considered a rare disease by the World Health Organization and has diverse causes, including genetic mutations, infections and metabolic disorders. Despite advancements in surgical management, patients remain at risk for complications such as amblyopia, strabismus and glaucoma. Visual outcomes heavily depend on early diagnosis, age at surgery, and adherence to postoperative care.

METHODS: Retrospective observational study including pediatric patients who underwent congenital cataract surgery at the Ophthalmology Department of Unidade Local de Saúde de Santo António in Porto, Portugal, between 2010 and 2023.

RESULTS: A total of 49 eyes (from 32 patients) with congenital cataract operated on a pediatric age were included. Most cases included bilateral cataracts (n= 34; 69.4%) and the most frequent location was posterior (n=22; 52.4%). The median age at surgery was 3.55 years old (0.06 – 16.90) and a family history of congenital cataract was positive in 4 (12.5%) patients. Laterality did not impact age at surgery (p=0.686). Primary intraocular lens (IOL) implantation was performed in 71.4% (n=35) of eyes, predominantly at older ages (5.81 (2.17-16.90) *vs* 0.30 (0.06-0.87); p<0.001). Patients submitted to surgery over 1 year of age had an improved final best-corrected visual acuity (p<0.001) and lower myopization rates across follow-up (p<0.001). A posterior Nd:YAG capsulotomy was required in 91.7% (n=11) of eyes not submitted to primary anterior vitrectomy/posterior capsulorhexis (n=12) at a median time of 4.04 years postoperatively and surgical reintervention was needed in 4 eyes. A diagnosis of glaucoma was made in 5 eyes (2 with persistent primary hyperplastic vitreous) at a median of 0.25 years postoperatively, with 3 eyes requiring glaucoma surgery. Additional variables significantly associated with an improved final BCVA were the absence of ocular structural anomalies and having bilateral cataracts.

CONCLUSION: Studying rare conditions like congenital cataracts is challenging due to the heterogeneous nature of real-life scenarios. However, documenting real-life management decisions and outcomes is crucial. Bilateral cataracts, being operated over the year of age and not having ocular structural anomalies, related to an improved final BCVA.

KEYWORDS: Cataract Extraction; Cataract/congenital; Child.

RESUMO

INTRODUÇÃO: A catarata congénita é considerada uma doença rara pela Organização Mundial da Saúde (OMS) e tem diversas causas, incluindo mutações genéticas, infeções e distúrbios metabólicos. Apesar dos avanços na sua gestão cirúrgica, continua a existir o risco de complicações como ambliopia, estrabismo e glaucoma. Os resultados visuais dependem sobretudo do diagnóstico precoce, da idade à data da cirurgia e da adesão aos cuidados pós-operatórios.

MÉTODOS: Estudo observacional retrospetivo incluindo doentes pediátricos submetidos a cirurgia de catarata congénita no Serviço de Oftalmologia da Unidade Local de Saúde de Santo António, Porto, Portugal, entre 2010 e 2023.

RESULTADOS: Foram incluídos um total de 49 olhos (de 32 doentes) com catarata congénita operados em idade pediátrica. A maioria dos casos apresentava cataratas bilaterais (n=34; 69,4%) e a localização mais frequente foi posterior (n=22; 52,4%). A idade média à data da cirurgia foi de 3,55 anos (0,06 – 16,90) e havia história familiar positiva em 4 (12,5%) doentes. A lateralidade não teve impacto na idade à data da cirurgia (p=0,686). O implante primário de lente intraocular (LIO) foi realizado em 71,4% (n=35) dos olhos, predominantemente em idades mais avançadas (5,81 (2,17-16,90) *vs* 0,30 (0,06-0,87); p<0,001). Doentes operados com mais de 1 ano de idade apresentaram uma melhor acuidade visual corrigida (AVC) final (p<0,001) e menores taxas de miopização ao longo do *follow-up* (p<0,001). Foi necessária capsulotomia posterior Nd:YAG em 91,7% (n=11) dos olhos não submetidos a vitrectomia anterior/capsulorrexis posterior primária (n=12), em média 4,04 anos após a cirurgia, e 4 olhos foram re-intervencionados. Cinco olhos foram diagnosticados com glaucoma (2 com persistência do vítreo primário hiperplástico), em média 0,25 anos após a cirurgia, sendo que 3 necessitaram de tratamento cirúrgico. Variáveis adicionais significativamente associadas a uma melhor AVC final foram a ausência de anomalias estruturais oculares e a presença de cataratas bilaterais.

CONCLUSÃO: Estudar doenças raras como a catarata congénita é desafiante devido à natureza heterogénea dos casos da vida real. Por esta razão é essencial documentar as decisões terapêuticas e os resultados na prática clínica. A presença de cataratas bilaterais, operadas após o 1º ano de vida e a ausência de anomalias estruturais oculares estão associadas a uma melhor AVC final.

PALAVRAS-CHAVE: Catarata Congénita; Criança; Extração de Catarata.

INTRODUCTION

Congenital cataract, which refers to an opacity of the lens detected at birth or at an early stage of childhood, is an important cause of visual disability worldwide, with an estimated incidence of 1.8 to 3.6 cases/10 000 children per year.[1] Its worldwide prevalence, estimated to be 4 to 5 cases/10 000 children, classifies it as a rare disease according to the World Health Organization (<6.5/10 000).[2] The etiology of congenital cataracts is diverse, with most bilateral cases involving mutations in genes encoding lens proteins.[3] Other causes include acquired intrauterine infections (especially congenital rubella syndrome), metabolic disorders, trauma, uveitis, corticosteroid use and radiation-induced cataracts.[3,4]

Although advancements in the surgical management of pediatric cataracts decreased the incidence of some postoperative complications, such as posterior capsule opacification,[5] these patients remain at risk for significant amblyopia, strabismus and glaucoma. Other postoperative complications include corneal edema, retinal hemorrhages/detachment, endophthalmitis, irregular pupil, iris heterochromia and visual axis opacities.[6]

Amblyopia occurs when there is deprivation of sensory information during sensitive periods of visual system development.[7] Thus, the visual prognosis depends critically on the age of cataract onset/diagnosis and surgery, as well as the laterality status, associated ocular/systemic conditions and adherence to postoperative optical correction and patching therapy.[8] The Infant Aphakia Treatment Study (IATS) demonstrated that children undergoing unilateral congenital cataract surgery can achieve good visual outcomes when early surgery is combined with consistent optical correction and part-time patching of the fellow eye.[9]

Strabismus can be a presenting symptom of visually significant cataracts. When present preoperatively, postoperative orthotropia is unlikely.[10] When reported following cataract surgery, strabismus is more likely to occur in unilateral cases compared to bilateral cases, with esotropia being more frequent.[10,11] The literature presents contradictory results regarding factors associated with an increased incidence of postoperative strabismus. These include the presence of nystagmus, a significant postoperative interocular best-corrected visual acuity (BCVA) difference and preservation of the posterior capsule, among others.[10,11] In

the IATS study, the incidence of strabismus in patients who underwent unilateral cataract surgery was unaffected by whether or not an intraocular lens (IOL) was implanted.[10]

Another late complication of pediatric cataract surgery is open-angle glaucoma, affecting up to 18% of infants operated on for a unilateral cataract.[12] The onset of glaucoma may occur up to years after surgery and for this reason, long-term follow-up is required. Its onset has been reported at a median of 4.3 years after surgery.[13] A younger age at surgery (especially before 4 weeks of age) is an important risk factor.12 Although primary IOL placement has long been considered protective, the IATS study did not identify a reduced glaucoma risk.[12,13]

The purpose of this retrospective observational study is to characterize a Portuguese congenital cataract patient cohort, its visual acuity outcomes and to study the association between congenital cataracts and strabismus, as well as the incidence of glaucoma. Investigating the real-life outcomes of rare and heterogeneous pathologies such as congenital cataracts is essential to improve our understanding of the disease and ultimately inform future clinical practices.

METHODS

This retrospective observational study used a pseudo-anonymized database of pediatric patients who underwent unilateral or bilateral cataract surgery at the Ophthalmology Department of Unidade Local de Saúde de Santo António in Porto, Portugal, between 2010 and 2023.

Collected data included age at cataract diagnosis and referencing motive, sex, ophthalmological/systemic/family history, cataract morphology and laterality status (unilateral versus bilateral), baseline BCVA and objective refraction, age at time of surgery, surgical technique performed, development of immediate/late complications (and management), need for reoperation and BCVA and objective refraction at 1-month, 1-year and at the last follow-up appointment. For children left aphakic and optically corrected with a contact lens, the refractive error at a vertex distance of 12 mm was calculated.

At our center, anterior vitrectomy is routinely carried out on all patients under six years of age undergoing congenital or juvenile cataract surgery. Primary posterior capsulorhexis is generally performed in children under eight years of age, but this will be adjusted depending on whether the child is considered cooperative for a subsequent Nd:YAG laser capsulotomy. Primary and secondary intraocular lens implantations are usually performed after 12 months of age.

Statistical analysis was performed using IBM SPSS Statistics 26. Normality of data was assessed using the Kolmogorov-Smirnov test. Our data analysis revealed a non-normal distribution for the studied variables. Categorical variables are presented as relative frequencies. Continuous variables are summarized as median and minimum and maximum values. A *p*-value inferior to 0.05 was considered statistically significant.

RESULTS

Out of a sample of 68 operated eyes, 19 were excluded due to the following reasons: traumatic cataract (8 eyes), uveitis-associated cataract (1 eye), metabolic-associated acquired cataract (2 eyes) and surgery performed only at adult age (8 eyes). As a result, 49 eyes of 32 patients with congenital cataract operated on at a pediatric age were included.

Patient demographics, as well as laterality, cataract type and preoperative BCVA and spherical equivalent (SE) are presented in Table 1. Most cases included bilateral cataracts (n= 34; 69.4%) and the most frequent type was posterior lamellar. The median age at surgery was 3.55 years old, with the oldest patient operated at 16.90 years of age. A family history of congenital cataract was positive in 4 (12.5%) patients and 3 of these had bilateral cataracts. A comprehensive evaluation of concomitant systemic and ophthalmological disease is displayed in Table 2, along with cataract laterality status. All patients with a preoperative exodeviation had unilateral cataracts, while only 16.7% of patients with a preoperative esodeviation had unilateral cataracts. Eyes with an exodeviation were operated on at an older age than those with an esodeviation (Table 1), but this difference was non-significant (p=0.069). Preoperative BCVA was also comparable between strabismus groups (p=0.486).

Table 1. Demographic data and clinical characteristics are displayed, including gender distribution, laterality, type of cataract, preoperative visual acuity and spherical equivalent, age at cataract surgery, age at last follow-up, follow-up duration, and family history.

Gender, *female patients* (%)	11 (34.4%)
Right eye (%)	23 (46.9%)
Laterality, *bilateral cataract* (%)	34 (69.4%)
Cataract type (%)	**n=42 (85.7%)**
Anterior	3 (7.1%)
Posterior	22 (52.4%)
Nuclear	8 (19.1%)
Total	9 (21.4%)
Strabismus, *patients*	12 (37.5%)
Esodeviation, *patients*	6 (18.75%)
Preoperative BCVA *(decimal)*	0.15 (0.01 - 0.50)
Esodeviation (1)	0.28 (0.01 - 0.50)
Exodeviation (2)	0.11 (0.01 - 0.20)
Preoperative BCVA (1 vs 2)	p=0.486
Preoperative SE (D)	0.00 (-10.00 - 5.00)
Age at surgery, *years*	3.55 (0.06 - 16.90)
Bilateral cataracts	3.55 (0.18 - 16.90)
Unilateral cataracts	3.79 (0.06 - 8.00)
Strabismus patients, age	2.18 (0.30 - 8.00)
Esodeviation (3)	2.18 (0.30 - 6.75)
Exodeviation (4)	7.00 (0.87 - 8.00)
Age at surgery (3 vs 4)	p=0.069
Age at last-follow-up, *years*	9.00 (1.86 - 26.40)
Follow-up time, *years*	474(0.04-12.66)
Family history, *patients* (%)	4 (12.5%)
Bilateral cataract patients with positive family history, *patients* (%)	3 (75.0%)

BCVA: bestcorrected visual acuity, SE: spherical equivalent, D: diopters.

Table 2. Ophthalmological and systemic associations in the study cohort are displayed, along with cataract laterality status for each group.

Ophthalmological associations, *patients*	
Esodeviation	6
Exodeviation	6
Persistent Hyperplastic Primary Vitreous	5
Posterior embriotoxon, iris stromal hypoplasia	1
Posterior lenticonus	1
Nystagmus	1
Unilateral cataract in,	
Esodeviation	1 (16.7%)
Exodeviation	6 (100%)
Persistent Hyperplastic Primary Vitreous	5 (100%)
Posterior embriotoxon, iris stromal hypoplasia	1 (100%)
Posterior lenticonus	1 (100%)
Nystagmus	0 (0%)
Systemic associations, *patients*	
Down Syndrome	2
Atopic dermatitis and food allergy	1
Mitochondrial disease	1
Demyelinating neuropathy and epilepsy	1
Gorlin Syndrome	1
Stickler Syndrome	1
Several congenital malformations	1
Bilateral cataract patients with positive systemic disease	6 (75.0%)

Laterality did not impact age at surgery (p=0.686), as illustrated in Fig. 1. This finding was consistent even when analyzing patients under 1 year of age (p=0.662). Primary

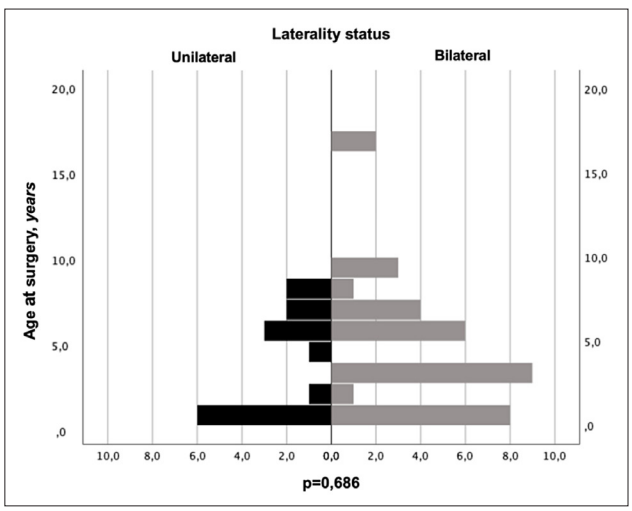

Figure 1. Double bar chart displaying the number of patients (x-axis) operated by age (y-axis) according to laterality status.

IOL implantation was performed in 71.4% (n=35) of eyes, predominantly at older ages (5.81 (2.17-16.90) vs aphakia at 0.30 (0.06-0.87); p<0.001) (Fig. 2 and Table 3). For eyes left aphakic (n=14), 4 underwent secondary IOL implantation at 1.5-years, 2-years and 6-years (2 eyes) postoperatively. Table 3 characterizes implanted IOLs and compares postoperative BCVA at 3 time points (1-month post-op, 1-year post-op and at last follow-up), SE and myopization rates between group 1 (primary IOL implantation) and group 2

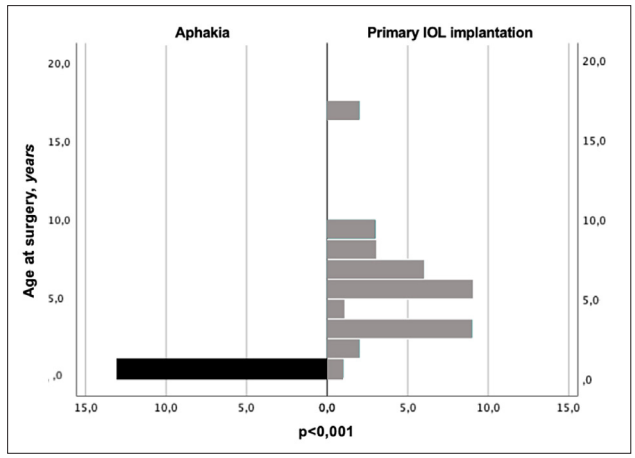

Figure 2. Double bar chart displaying the number of patients (x-axis) operated by age (y-axis) according to the chosen management strategy.

Table 3. Summary of postoperative visual and refractive outcomes for patients undergoing primary IOL implantation (group 1) versus those left aphakic (group 2). The table displays BCVA and SE in 3 time points, as well as myopization rates for both groups. The statistical significance for the differences in final BCVA and myopization rates between groups is displayed.

Primary IOL implantation (group 1)	**n=35 (71.4%)**
Age at surgery, *years*	5.81 (2.17-16.90)
Axial length (mm)	21.96 (18.17 - 32.40)
IOL power (D)	23.00 (1.00 - 32.00)
IOL power target (SRK/T) (D)	0.55 (-1.50 - 2.00)
1-month postop BCVA *(decimal)*	0.50 (0.01 - 1.00)
1-year postop BCVA *(decimal)*	0.60 (0.01 - 1.00)
Last follow-up BCVA *(decimal)*	0.70 (0.01 - 1.00)
1-month SE (D)	0.50 (-1.75 - 3.25)
1-year SE (D)	0.00 (-2.50 - 4.75)
Last follow-up SE (D)	-1.00 (-4.50 - 2.25)
Myopization (D) 1 year postop	0,00 (-2.25-1.50)
Myopization (D/year) total follow-up	-0.17 (-0.86-0.12)
Aphakia (group 2)	**n=14 (28.6%)**
Age at surgery, *years*	0.30 (0.06-0.87)
1-month postop BCVA *(decimal)*	--
1-year postop BCVA *(decimal)*	--
Last follow-up BCVA *(decimal)*	0.07 (0.00 - 0.50)
1-month SE (D)	18.50 (11.00 - 27.00)
1-year SE (D)	13.25 (6.00 - 20.00)
Last follow-up SE (D)	4.88 (-1.25 - 10.50)
Myopization (D) 1 year post op	-4.75 (-14.00 - -0.50)
Myopization (D/year) total follow-up	-2.39 (-6.34 - -0.90)
Last follow-up BCVA *(decimal)* (group 1 *vs* group 2)	p<0.001
Myopization (D/year) 1 year post op (group 1 *vs* group 2)	p<0.001
Myopization (D/year) total follow-up (group 1 *vs* group 2)	p<0.001

BCVA: bestcorrected visual acuity, SE: spherical equivalent, D: diopters.

(primary aphakia). The aphakia group had a higher myopization rate throughout the follow-up period (p<0.001) and worse final BCVA (p<0.001).

A posterior Nd:YAG capsulotomy was required in 91.7% (n=11) of eyes not submitted to primary anterior vitrectomy/

posterior capsulorhexis (n=12) at a median time of 4.04 years postoperatively (4.04 (0.68 – 9.51)). All had been implanted with an IOL and were over the age of 5 years (which is in line with the usual practice at our center), except for a child with Stickler syndrome, operated on at the age of 4. A surgical re-intervention was needed in 4 eyes due to vitreous strands to the main incision (1 eye), excessive inflammation requiring lens explantation (1 eye) and bilateral anterior chamber IOL subluxation (of a secondarily implanted IOL). A diagnosis of glaucoma was made in 5 eyes (2 from the same patient) at a median of 0,25 years postoperatively (0.23 – 3.64). Of these, 2 had been submitted to cataract surgery with primary IOL implantation and 3 had been left aphakic, at a median age at surgery of 0,18 years (0.06 – 3.56). Two of these eyes also had persistent fetal vasculature (PFV) and the other 2 had postoperative complications requiring early reintervention. Subsequent glaucoma surgery was needed in 3 eyes (cyclophotocoagulation and deep sclerotomy) and hypotensive topical drugs were being applied to 4 (80%) eyes at the end of follow-up. Fig. 3 displays the evaluation of different variables (age at surgery, surgical management strategy, gender, presence/absence of strabismus, laterality, location and presence of structural anomalies) in association with final BCVA. Variables significantly associated with an improved BCVA was being over 1 year of age at the time of surgery (p<0.001), the decision to primarily implant an IOL (p<0.001), having bilateral cataracts (p=0.007) and the absence of structural anomalies (p<0.001). Structural anomalies considered for the analysis included eyes described in Table 2 (*Ophthalmological associations*), excluding strabismus and nystagmus.

DISCUSSION

In our study, most patients (65.6%) were male, which is in line with male-to-female incidence ratios reported in other studies.[14,15] Bilateral cataracts were also more common, but laterality status did not significantly impact age at surgery, even under the first year of age. The most common cataract type was posterior lamellar. These include opacities of posterior layers of the lens, often with opacification of the adjacent cortex,[6] carrying a worse visual prognosis in comparison to anterior polar/nuclear cataracts.[16] This likely explains why over half of all operated eyes in our sample had this type of cataract. Since genetic mutations account for the majority of cases of bilateral cataracts,[3] with both dominant and recessive inheritance patterns,[3,17] it is unsurprising that most patients with positive family history in our sample had bilateral cataracts. The systemic conditions described in this cohort had already been associated with cataract development, namely for Down, Stickler and Gorlin syndromes and mitochondrial diseases.[16,18,19] Atopic dermatitis has also been associated with an increased risk, especially in severe forms of the disease.[20]

Regarding ophthalmological associations, preoperative strabismus included only horizontal deviations. The incidence of preoperative strabismus was 37.5%, slightly higher than IATS reported incidence.[10] The patients enrolled in the IATS study had a median age of 1.8 months,[21] which limits its comparability with our study. The number of patients with esotropia and exotropia was equal, though esotropia is more commonly reported in the literature.[22] A unilateral cataract was more commonly associated with an exodeviation and bilateral cataracts with esodeviations. The age of onset of vision loss, interorbital distance, and refractive error in the unaffected/less affected eye are known factors in determining the direction of sensory strabismus.[23,24] In our sample, we observed a tendency for eyes with an exodeviation to be operated on at an older age than those with an esodeviation, although their preoperative BCVA was comparable. This finding is in line with the observation that most patients with very early visual loss develop an esotropia, whereas those who experience visual loss later tend to develop an exotropia. Although reported incidences of strabismus after cataract surgery show esotropia to outnumber exotropia,[10,25] we only assessed preoperative strabismus and did not evaluate it prospectively.

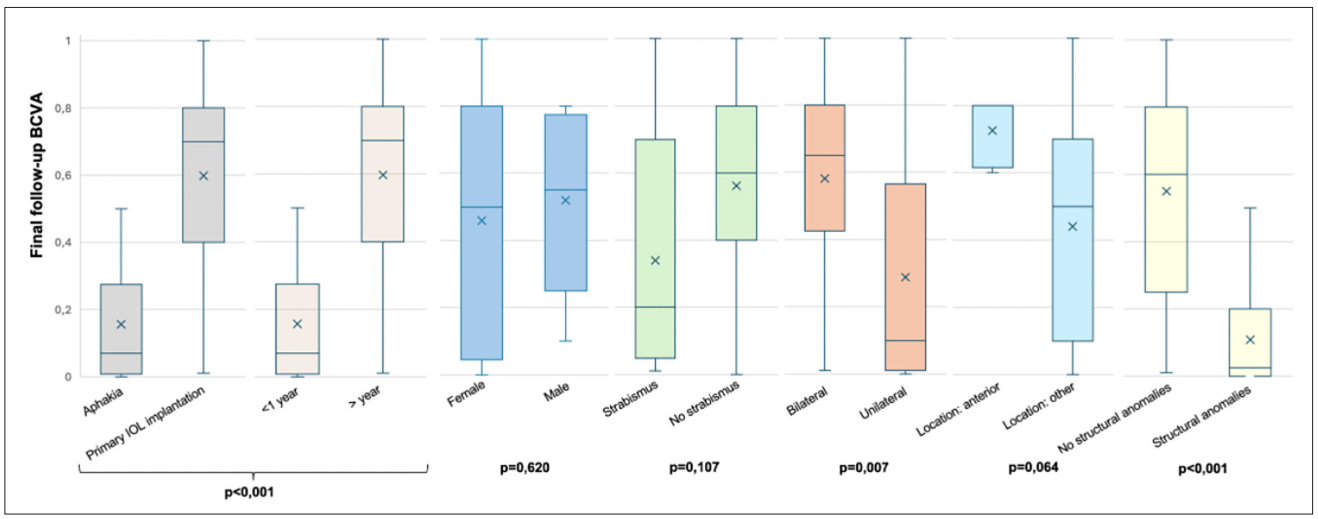

Figure 3. A clustered boxplot of potential visual prognostic factor is shown. The p-value for each variable is shown under the x-axis.

Regarding the surgical management, our results are in accordance with the literature. No patient was left aphakic when operated on over 1 year of age. In the IATS study, including only unilateral congenital cataracts, the mean grating visual acuity (at 12 months of age), median optotype (HOTV test) BCVA (at 4.5 years of age) and median logMAR (ETDRS) BCVA (at 10.5 years of age)[26] did not differ significantly between surgical strategies (primary IOL implantation vs aphakia) in patients operated in the first 6 months of life.[27,28] Since there was a higher incidence of adverse events in the primary IOL implantation group, specifically iris prolapse during IOL placement,[27] lens reproliferation into the visual axis, pupillary membrane formation, and corectopia, a recommendation was drawn to leave aphakic an eye operated on an infant younger than 7 months of age.[27,28] Only 4 of the 14 patients left aphakic in our cohort were submitted to a secondary IOL implant during follow-up, at 1.5 (1 eye), 2 (1 eye) and 6 years-postoperatively (2 eyes). Additionally, the surgical strategy in our cohort did impact final BCVA, with primary IOL implantation patients (>1 year of age at time of surgery) exhibiting an improved final BCVA. Age and primary IOL implantation are correlated and their impact on the final BCVA is indistinguishable in our case series. Understandably, patients operated at an older age (with IOL placement) most likely had less visually significant cataracts, hence a better prognosis. When predicting the optimal IOL power, a myopic shift (with growth) needs to be anticipated, and so an undercorrection is usually applied, which is in line with our calculated target 0.55D (-1.50 – 2.00)D. Our minimum chosen target was -1.50D for an 8 year-old patient who did not tolerate using optical correction preoperatively. Of note, our sample has no planned undercorrections over +2.00D. This could be explained with Fig. 2, since these patients were operated mostly over 1 year of age (median age 5.81 (2.17-16.90)), where typically applied overcorrections are lower.[29] Regarding the myopization rate, it was significantly higher in eyes left aphakic in comparison to eyes implanted primarily with an IOL. An important point is that patients were on average older in the primary implantation group, so the myopization rates would be expectingly lower. Regarding aphakic patients, the IATS study found an average myopization rate of -2.11D/year from surgery to 18 months post-randomization (PR), -0.68D/year from 1.5 years to 5 years PR and −0.35/year from 5 to 10 years PR.[30] In the first year after surgery, myopization rates in our aphakia group were higher than reported (-4.75 (-14.00 – +0.50))D/year. A patient with unilateral congenital cataract and persistent hyperplastic primary vitreous experienced the maximum myopic shift of -14.00D within 1 year and was diagnosed with glaucoma four years after surgery. Considering postoperative complications, a posterior Nd:YAG capsulotomy was required for almost all (91.7%; n=11) eyes that had not been submitted to a primary anterior vitrectomy/posterior capsulorhexis. Given the varied timing until capsulotomy (median of 4.04 years (0.68 – 9.51)), we emphasize the need for long-term monitoring.

Regarding glaucoma, 10.20% of eyes were affected at a median of 0.25 years postoperatively (0.23 – 3.64), a lower incidence compared to the IATS study (18% by 5 years of age)[12] and sooner after cataract surgery than reported (mean of 4 to 5 years after cataract removal).[12] Reported incidences of glaucoma differ greatly in the literature due to different follow-up times (hindering detection), glaucoma definitions (hindering inclusion) and study populations.[12,31,32] The potential protective effect of primary IOL implantation in preventing glaucoma after cataract removal in infancy is debated, but the IATS did not support this protective effect.[12] Our number of glaucoma cases (n=5) is small to conclude this topic, especially because there is a similar number of cases in each surgery group. Patients were relatively young at cataract removal (0.18 years (0.06 – 3.56)), and this is a risk factor for glaucoma both in the 1-year and 5-year results of the IATS.[12,33] Specifically, age ranges conferring increased risk were 28-48 days-old *versus* 49-210 days-old.[12,33] Two of our patients had PFV, another known risk factor for glaucoma that has been found to increase the risk 3.1 times.[33]

Variables significantly associated with an improved BCVA were being over 1 year of age at the time of surgery, the decision to primarily implant an IOL, having bilateral cataracts and the absence of ocular structural anomalies. In the literature, visual prognostic factors are highly variable. Factors linked to a poorer postoperative visual acuity have included male gender, presence of nonophthalmic disorders, preoperative nystagmus, age at surgery < 1 year, aphakia, postoperative complications and being of non-Caucasian ethnicity.[34,35] Another important factor to be considered is compliance with postoperative occlusion. This was not evaluated in this study, but others have found it to be the factor most strongly associated with the visual outcome of both unilateral and bilateral cataracts.34 Our results regarding the effect of age (and subsequently the decision to primarily implant an IOL) in final BCVA are according to the literature, as well as the effect of ocular structural anomalies (in our sample mostly persistent hyperplastic primary vitreous), whose prognosis depends on the specific pathology involved. In our sample, bilateral cataracts had a better visual prognosis. Since the laterality status (unilateral *vs* bilateral) did not differ according to the age at surgery, we can attribute this effect to the competitive inhibition (pronounced in unilateral cataracts), alongside the impact of the stimulus deprivation itself.

Two major limitations of this study are its retrospective, non-interventional nature and the relatively small sample size, which limits the generalizability of results. Patients were also included irrespective of the follow-up duration, with some having ceased follow-up at our center. Shorter follow-up periods may not be sufficient to identify late-onset postoperative complications, such as glaucoma or retinal detachment.

Although studying rare and heterogeneous conditions like congenital cataracts presents numerous challenges, particularly in terms of homogenizing data to draw unbiased conclusions, it is crucial to document real-life management and outcomes. Only by doing so can we truly understand the effectiveness of our various treatment approaches and enhance the care we provide to our patients.

DECLARAÇÃO DE CONTRIBUIÇÃO / CONTRIBUTORSHIP STATEMENT

FB: Redação do manuscrito.

FB, BR: Responsáveis pela colheita de dados e amostras, apresentação dos resultados e revisão do manuscrito.

JHM, LM, FC, SM, VM, RP, PM: Responsáveis pela orientação clínica dos doentes,colheita de amostrase revisão do manuscrito.

BR, JHM, LM, FC, SM, VM, RP, PM: Conceção, desenho do estudo, revisão do manuscrito, supervisão do projeto e contribuição com expertise para a sua conclusão.

Todos os autores leram e aprovaram o manuscrito final.

FB: Writing the manuscript.

FB, BR: Responsible for collecting data and samples, presenting the results and manuscript revision.

JHM, LM, FC, SM, VM, RP, PM: Responsible for clinical guidance of patients, sample collection and manuscript revision.

BR, JHM, LM, FC, SM, VM, RP, PM: Conception, design of the study, review of the manuscript, supervision of the project and contribution of expertise to its completion.

All authors: read and approved the final manuscript.

RESPONSABILIDADES ÉTICAS

Conflitos de Interesse: Os autores declaram a inexistência de conflitos de interesse na realização do presente trabalho.

Fontes de Financiamento: Não existiram fontes externas de financiamento para a realização deste artigo.

Confidencialidade dos Dados: Os autores declaram ter seguido os protocolos da sua instituição acerca da publicação dos dados de doentes.

Proteção de Pessoas e Animais: Os autores declaram que os procedimentos seguidos estavam de acordo com os regulamentos estabelecidos pela Comissão de Ética responsável e de acordo com a Declaração de Helsínquia revista em 2024 e da Associação Médica Mundial.

Proveniência e Revisão por Pares: Não comissionado; revisão externa por pares.

ETHICAL DISCLOSURES

Conflicts of Interest: The authors have no conflicts of interest to declare.

Financing Support: This work has not received any contribution, grant or scholarship

Confidentiality of Data: The authors declare that they have followed the protocols of their work center on the publication of patient data.

Protection of Human and Animal Subjects: The authors declare that the procedures followed were in accordance with the regulations of the relevant clinical research ethics committee and those of the Code of Ethics of the World Medical Association (Declaration of Helsinki as revised in 2024).

Provenance and Peer Review: Not commissioned; externally peer-reviewed.

REFERENCES

1. Sheeladevi S, Lawrenson JG, Fielder AR, Suttle CM. Global prevalence of childhood cataract: a systematic review. Eye. 2016;30:1160-9. doi: 10.1038/eye.2016.156.
2. Widdus R. Public-private partnerships for health: their main targets, their diversity, and their future directions. Bull World Health Organ. 2001;79:713-20.
3. Bell SJ, Oluonye N, Harding P, Moosajee M. Congenital cataract: a guide to genetic and clinical management. Ther Adv Rare Dis. 2020;1:2633004020938061. doi: 10.1177/2633004020938061.
4. Tătaru CI, Tătaru CP, Costache A, Boruga O, Zemba M, Ciuluvică RC, et al. Congenital cataract - clinical and morphological aspects. Rom J Morphol Embryol. 2020;61:105-12. doi: 10.47162/RJME.61.1.11.
5. Nihalani BR, Vanderveen DK. Technological advances in pediatric cataract surgery. Semin Ophthalmol. 2010;25:271-4. doi: 10.3109/08820538.2010.518836.
6. Lambert SR, Lyons CJ. Taylor and Hoyt's Pediatric Ophthalmology and Strabismus. 5th ed. Amsterdam: Elsevier;2016.
7. Feng Y, Collignon O, Maurer D, Yao K, Gao X. Brief Postnatal Visual Deprivation Triggers Long-Lasting Interactive Structural and Functional Reorganization of the Human Cortex. Front Med. 2021;8:752021. doi: 10.3389/fmed.2021.752021.
8. Oshika T, Endo T, Kurosaka D, Matsuki N, Miyagi M, Mori T, et al. Long-term surgical outcomes of pediatric cataract-multivariate analysis of prognostic factors. Sci Rep. 2023;13:21645. doi: 10.1038/s41598-023-49166-2.
9. Drews-Botsch C, Celano M, Cotsonis G, Hartmann EE, Lambert SR; Infant Aphakia Treatment Study Group. Association Between Occlusion Therapy and Optotype Visual Acuity in Children Using Data From the Infant Aphakia Treatment Study: A Secondary Analysis of a Randomized Clinical Trial. JAMA Ophthalmol. 2016;134:863-9. doi: 10.1001/jamaophthalmol.2016.1365.
10. Bothun ED, Lynn MJ, Christiansen SP, Neely DE, Vanderveen DK, Kruger SJ, et al. Sensorimotor outcomes by age 5 years after monocular cataract surgery in the Infant Aphakia Treatment Study (IATS). J AAPOS. 2016;20:49-53. doi: 10.1016/j.jaapos.2015.11.002.
11. Lee SJ, Kim WS. Factors associated with strabismus after cataract extraction and primary intraocular lens implantation in congenital cataracts. Int J Ophthalmol. 2014;7:522-7. doi: 10.3980/j.issn.2222-
12. Freedman SF, Lynn MJ, Beck AD, Bothun ED, Örge FH, Lambert SR, et al. Glaucoma-Related Adverse Events in the First 5 Years After Unilateral Cataract Removal in the Infant Aphakia Treatment Study. JAMA Ophthalmol. 2015;133:907-14. doi: 10.1001/jamaophthalmol.2015.1329.
13. Mataftsi A, Haidich AB, Kokkali S, Rabiah PK, Birch E, Stager DR Jr, et al. Postoperative glaucoma following infantile cataract surgery: an individual patient data meta-analysis. JAMA Ophthalmol. 2014;132:1059-67. doi: 10.1001/jamaophthalmol.2014.1042..
14. Sharma AK, Shah DN, Upadhyay MP, Thapa M. Demography and Etiology of Congenital Cataract in a Tertiary Eye Centre of Kathmandu, Nepal. Health Renaissance. 2015; 12.doi: 10.3126/hren.v12i1.11975.
15. Katibeh M, Eskandari A, Yaseri M, Hosseini S, Ziaei H. The

gender issue in congenital and developmental cataract surgery. J Ophthalmic Vis Res. 2013;8:308-13.
16. Amaya L, Taylor D, Russell-Eggitt I, Nischal KK, Lengyel D. The morphology and natural history of childhood cataracts. Surv Ophthalmol. 2003;48:125-44. doi: 10.1016/s0039-6257(02)00462-9.
17. Patel A, Hayward JD, Tailor V, Nyanhete R, Ahlfors H, Gabriel C, et al. The Oculome Panel Test: Next-Generation Sequencing to Diagnose a Diverse Range of Genetic Developmental Eye Disorders. Ophthalmology. 2019;126:888-907. doi: 10.1016/j.ophtha.2018.12.050.
18. Snead MP, McNinch AM, Poulson AV, Bearcroft P, Silverman B, Gomersall P, et al. Stickler syndrome, ocular-only variants and a key diagnostic role for the ophthalmologist. Eye. 2011;25:1389-400. doi: 10.1038/eye.2011.201.
19. Moramarco A, Himmelblau E, Miraglia E, Mallone F, Roberti V, Franzone F, et al. Ocular manifestations in Gorlin-Goltz syndrome. Orphanet J Rare Dis. 2019;14:218. doi: 10.1186/s13023-019-1190-6.
20. Jeon HS, Choi M, Byun SJ, Hyon JY, Park KH, Park SJ. Association of Pediatric Atopic Dermatitis and Cataract Development and Surgery. JAMA Ophthalmol. 2018;136:912-8. doi: 10.1001/jamaophthalmol.2018.2166.
21. Infant Aphakia Treatment Study Group; Lambert SR, Buckley EG, Drews-Botsch C, DuBois L, Hartmann E, et al. The infant aphakia treatment study: design and clinical measures at enrollment. Arch Ophthalmol. 2010;128:21-7. doi: 10.1001/archophthalmol.2009.350.
22. David R, Davelman J, Mechoulam H, Cohen E, Karshai I, Anteby I. Strabismus developing after unilateral and bilateral cataract surgery in children. Eye. 2016;30:1210-4. doi: 10.1038/eye.2016.162.
23. Kim IG, Park JM, Lee SJ. Factors associated with the direction of ocular deviation in sensory horizontal strabismus and unilateral organic ocular problems. Korean J Ophthalmol. 2012;26:199-202. doi: 10.3341/kjo.2012.26.3.199.
24. Havertape SA, Cruz OA, Chu FC. Sensory strabismus - Eso or exo? J Pediatr Ophthalmol Strabismus. 2001;38:327-30; quiz 354-5. doi: 10.3928/0191-3913-20011101-05..
25. France TD, Frank JW. The association of strabismus and aphakia in children. J Pediatr Ophthalmol Strabismus. 1984;21:223-6. doi: 10.3928/0191-3913-19841101-06.
26. Lambert SR, Cotsonis G, DuBois L, Nizam Ms A, Kruger SJ, Hartmann EE, et al. Long-term Effect of Intraocular Lens vs Contact Lens Correction on Visual Acuity After Cataract Surgery During Infancy: A Randomized Clinical Trial. JAMA Ophthalmol. 2020;138:365-72. doi: 10.1001/jamaophthalmol.2020.0006.
27. Infant Aphakia Treatment Study Group; Lambert SR, Lynn MJ, Hartmann EE, DuBois L, Drews-Botsch C, et al. Comparison of contact lens and intraocular lens correction of monocular aphakia during infancy: a randomized clinical trial of HOTV optotype acuity at age 4.5 years and clinical findings at age 5 years. JAMA Ophthalmol. 2014;132:676-82. doi: 10.1001/jamaophthalmol.2014.531.
28. Infant Aphakia Treatment Study Group; Lambert SR, Buckley EG, Drews-Botsch C, DuBois L, Hartmann EE, et al. A randomized clinical trial comparing contact lens with intraocular lens correction of monocular aphakia during infancy: grating acuity and adverse events at age 1 year. Arch Ophthalmol. 2010;128:810-8. doi: 10.1001/archophthalmol.2010.101.
29. Touzé R, Dureau P, Edelson C, Borella Y, Barjol A, de Laage de Meux P, et al. Congenital cataract surgery: long-term refractive outcomes of a new intraocular lens power correction formula. Acta Ophthalmol. 2022;100:e1641-5. doi: 10.1111/aos.15134.
30. Lambert SR, Nizam A, DuBois L, Cotsonis G, Weakley DR Jr, Wilson ME, et al. The Myopic Shift in Aphakic Eyes in the Infant Aphakia Treatment Study After 10 Years of Follow-up. Eye Contact Lens. 2021;47:108-12. doi: 10.1097/ICL.0000000000000718.
31. Egbert JE, Christiansen SP, Wright MM, Young TL, Summers CG. The natural history of glaucoma and ocular hypertension after pediatric cataract surgery. J AAPOS. 2006;10:54-7. doi: 10.1016/j.jaapos.2005.07.002.
32. Lambert SR, Purohit A, Superak HM, Lynn MJ, Beck AD. Long-term risk of glaucoma after congenital cataract surgery. Am J Ophthalmol. 2013;156:355-361.e2. doi: 10.1016/j.ajo.2013.03.013.
33. Beck AD, Freedman SF, Lynn MJ, Bothun E, Neely DE, Lambert SR, et al. Glaucoma-related adverse events in the Infant Aphakia Treatment Study: 1-year results. Arch Ophthalmol. 2012;130:300-5. doi: 10.1001/archophthalmol.2011.347.
34. Chak M, Wade A, Rahi JS. Long-term visual acuity and its predictors after surgery for congenital cataract: Findings of the British Congenital Cataract Study. Invest Ophthalmol Vis Sci. 2006;47:4262-9. doi: 10.1167/iovs.05-1160.
35. Bonaparte LA, Trivedi RH, Ramakrishnan V, Wilson ME. Visual acuity and its predictors after surgery for bilateral cataracts in children. Eye. 2016;30:1229-33. doi: 10.1038/eye.2016.166.

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