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Clinical manifestation of congenital aniridia in Ukrainian children and adolescents

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Purpose: *To investigate clinical manifestations of congenital aniridia (CA) in Ukrainian children and adolescents.*

Material and Methods: *Twenty four children and adolescents (47 eyes; group 1 of children under 11 years and group 2 of adolescents aged 11 to 18 years) and 5 adult parents (age, 38 to 48 years; group 3) of patients with CA were included in the study. Of these, 72.4% had familial aniridia and 27.6%, sporadic aniridia. The eye examination included gonioscopy, keratometry, ultrasound biometry, ultrasound examination of the anterior and posterior segments, phosphene assessment, and optical computed tomography.*

Results: *Most patients (75.0% in group 1, 86.7% in group 2, and 100% in group 3) had biomicroscopic evidence of complete aniridia. Aniridia-associated keratopathy (AAK) was found in all groups, with differences in clinical manifestations and severity of AAK between groups. Rates of stages A, B, C and D AAK were 0%, 12.5%, 0%, and 0%, respectively, for group 1, 0%, 40%, 20%, and 13.3%, respectively, for group 2, and 0%, 0%, 70%, and 30%, respectively, for group 3. Congenital cataract was found in 81.3% of eyes in group 1, and progressive cataract, in 86.7% of eyes in group 2. The rate of congenital glaucoma with corneal edema and buphthalmos was 12.5% in group 1. The rate of secondary glaucoma in groups 2 and 3 was 60%. Congenital foveal hypoplasia was observed in 93.75% and 93.3% of the eyes in children and adolescents, respectively. In groups 1, 2 and 3, visual acuity was more commonly ≤ 0.1, and 0.1-0.25 in 37.5%, 26.7%, and 20.0% of eyes, respectively.*

Conclusion: *CA is a rare bilateral panocular genetic disorder, which in 93.3-93.8% of our cases was associated with foveal hypoplasia, causing low vision. Clinical manifestations of CA (AAK, glaucoma, cataract, foveal hypoplasia, nystagmus, etc.) were found to increase in severity with age. Parents of aniridic children exhibited the worst state of the eye. Clinical manifestations of CA were more severe, with higher rates of complications, in eyes with familial CA, compared to eyes with sporadic CA.*

Keywords:

congenital aniridia, aniridia-associated keratopathy, cararact, nystagmus

Introduction

Congenital aniridia (CA) is a rare ophthalmic genetic disorder characterized by partial or total loss of iris tissue and frequently defined by hypoplasia of iris. It was first described by the Italian researcher Barrata in 1818 [1, 2]. The condition is bilateral in 98% of all affected patients [3]. The degree of iris hypoplasia varies significantly between patients with iris tissue only observed by biomiscroscopy, gonioscopy or optical coherence tomography (OCT) of the anterior segment. The prevalence of CA ranges from 1:40,000 to 1:100,000. No racial or sexual differences are recognized [4, 5]. CA is inherited in an autosomal dominant manner due to a mutation in the PAX6 gene. That means that a child only needs one copy of the mutated gene, from either biological parent, to be affected by the disease. Autosomal dominant aniridia accounts for 30-50% of cases. About 13% to 33% of people with aniridia have sporadic aniridia. This variant of aniridia correlates with nephroblastoma (Wilms tumor) as a part of WAGR syndrome (Wilms tumor, aniridia, genitourinary anomalies, and mental retardation). Out of the patients with CA, 5% had WAGR syndrome [6] and 2% had Gillespie syndrome [7]. The ocular abnormalities associated with aniridia include foveal and optic nerve hypoplasia, cataract, glaucoma and keratopathy [8]. Studies on manifestations of CA are important because the disease is rare, has a variety of clinical manifestations, and significantly affects vision.

The purpose of the study was to investigate clinical manifestations of CA in Ukrainian children and adolescents.

Material and Methods

This prospective study followed the tenets of the Declaration of Helsinki, and the study protocol was approved by the institutional review board of the institute (Date of Approval: 29.07.2025). Written informed consent was obtained from all adult patients and all parents of child or adolescent patients. Twenty four aniridic children and adolescents (47 eyes; age range, 5 months to 18 years)

and 5 aniridic adult parents (10 eyes; age range, 38-48 years) of patients under 18 years underwent examination at the Pediatric Ophthalmology Department. None of the subjects exhibited any systemic manifestations of WAGR syndrome or Gillespie syndrome during the study period.

According to the Convention on The Right of the Child, children are individuals under the age of 18 years. The United Nations (UN) defines adolescents as individuals being 10-19 years old [9].

Aniridic individuals and eyes were divided into three groups depending on age. Group 1 (children 11 years or younger) comprised 16 individuals and 32 eyes (56.1%). Group 2 (adolescents 11 to 18 years) comprised 8 individuals and 15 eyes (26.3%). Group 3 (parents of pediatric patients) comprised 5 individuals and 10 eyes (17.6%). Familial aniridia was determined based on family history of aniridia. Of the 29 subjects, 21 (72.4%) had familial aniridia and 8 (27.6%) had sporadic aniridia, and the latter was found only among children under 11 years. Corneal changes in aniridia were classified according to the system proposed by Mackman [10].

The eye examination included biomicroscopy, gonioscopy, detailed ophthalmoscopy, tonometry, tonography, refractometry, keratometry, ultrasound biometry, ultrasound examination of the anterior and posterior segments, phosphene assessment, and OCT. Eyes of small-age children were examined under general anesthesia.

JASP software (JASP team, version 0.19.2, for Windows, Amsterdam Uni-versity, Amsterdam, Netherlands) was employed for statistical analysis. Categori-cal data are presented as frequencies (n) and percentage (%).The Shapiro–Wilk test was used to test for normality. Numerical data are presented as median (Me) with interquartile range (IQR). Chi-square or Mann-Whitney U tests were used for comparison between groups. P values < 0.05 were considered significant.

Results

Photophobia was the major complaint in all patients. Consequently, all of them displayed characteristic facial features (“narrowed eyes”, frowning, and partial ptosis) as involuntary measures of light protection (Fig. 1).

Most patients (75.0% in group 1, 86.7% in group 2, and 100% in group 3) had biomicroscopic evidence of complete aniridia (Fig. 2A). A quarter of children in group 1 and 13.3% of adolescents in group 2 had biomicroscopic evidence of incomplete aniridia with the preservation of a 0.5 to 1.0-mm-wide band of the peripheral iris (Fig. 2B).

Aniridia-associated keratopathy (AAK) was found in all age groups, but there was a difference between groups in terms of clinical manifestations and Mackman stages (Table 1).

Corneal transparency was preserved in most children. Only 4 eyes (12.5%) in two children exhibited mild (stage B) peripheral AAK, with ≤ 3.0-mm corneal penetration by the limbal pannus and preservation of central corneal transparency. There was evidence of corneal edema in both eyes in two children with congenital glaucoma (Table 1). Eyes of adolescent patients exhibited deterioration of the cornea. Of these eyes, 40.0% showed stage B peripheral AAK and conjunctival ingrowth into the peripheral and paracentral cornea (Fig. 3A), 20.0% showed stage C AAK involving the central cornea and apparent corneal vascularization (Fig. 3B), and 13.5% (2) showed stage D AAK. Of the eyes of adult patients, 70% exhibited stage C AAK, and 30%, stage D AAK (Fig. 4) with complete corneal conjunctivalization and vascularization.

Fig. 5 shows the presence of AAK depending on the age, with a significant difference between children under 11 years and adolescents of 11 to 18 years (1) and children under 11 years and adult parents (2).

Congenital cataract was diagnosed in 81.3% of eyes of children with CA. In the majority of cases, it was partial (an anterior polar or capsular cataract), with preservation of a significant portion of transparent lens (Fig. 6A). Grade 1 subluxation of a transparent lens was present in 2 cases (12.5%). Of the eyes of adolescent patients, 86.7% had progressive lamellar cataract (Fig. 6B) or partially resolved cataract (Fig. 6C) with calcification. Additionally, superior lens subluxation was observed in 33.3% of these eyes. Three adult parents received cataract surgery (three pseudophakic and three aphakic eyes). Four eyes in two adult parents had progressive lamellar cataract (Table 1).

Congenital glaucoma with corneal edema and buphthalmos was found in 12.5% (Fig. 7) of cases, and was an indication for urgent surgery. Of the eyes of adolescent and adult patients, 60% had secondary glaucoma, which in the

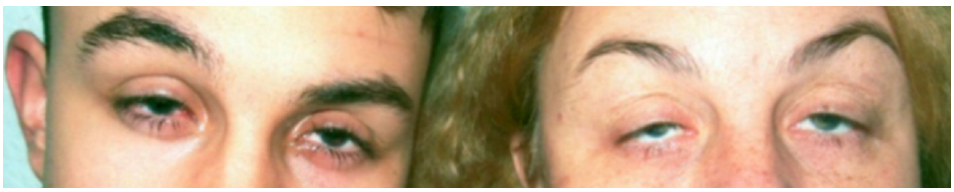


Fig. 1. A 15-year-old adolescent and his mother with congenital binocular aniridia and partial ptosis (more apparent in the mother).

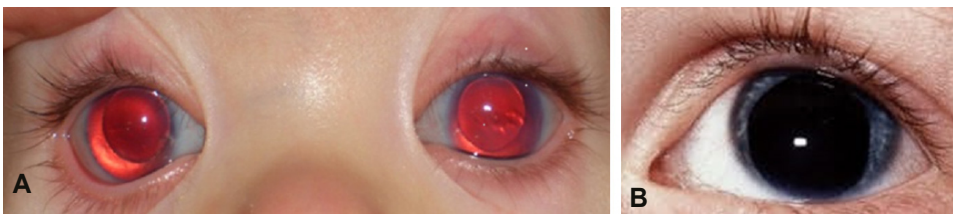


Fig. 2. (A): Congenital binocular complete aniridia with superior subluxation of transparent lenses in a 12-month child. (B): Congenital partial aniridia in a 12-year-old adolescent.

Table 1. Clinical manifestations of congenital aniridia and visual functions in patients of the study groups

Clinical manifestations	Children under 11 years	Adolescents aged 11 to 18 years	Adults (parents)
	16 children (32 eyes) eyes/ percentage	8 adolescents (15 eyes) eyes / percentage	5 adults (10 eyes) eyes/ percentage
Aniridia (on biomiscrosopy):			
complete	24 / 75.0	13 / 86.7	10 / 100.0
incomplete	8 / 25.0	2 / 13.3	-
Anterior microphthalmia	4	-	2
Buphthalmos	4	7	-
Megalocornea	4	-	-
Aniridia-associated keratopathy:	4 / 12.5 [a,b]	11 / 73.3	10 / 100.0
stage B	4 / 12.5 [a]	6 / 40.0 [c]	-
stage C	-	3 / 20.0	7 / 70.0
stage D	-	2 / 13.3	3 / 30.0
Corneal edema	2	3	
Persistent pupillary membrane	10 [a]	-	-
Cataract:			
Congenital (anterior polar or anterior capsular)	26 / 81.3 [a,b]	-	-
Progressive (semiresolved, lamellar or calcified)	0 [a,b]	13 / 86.7	4 / 40.0
Superior lens subluxation	2 / 6.25	5 / 33.3	4 / 40.0
Aphakia	-	-	3
Pseudophakia	-	2	3
Glaucoma:	4 / 12.5 [a,b]		
Congenital	4 / 12.5%	-	-
Secondary	0 [a,b]	9 / 60.0	6 / 60.0
Foveolar hypoplasia	30 / 93.75	14 / 93.3	6 / 60.0
Optic nerve hypoplasia and partial optic atrophy	8 / 25.0 [a]	10 / 66.7 [c]	-
Nystagmus	28 / 87.5	13 / 86.7	10 / 100.0
Refractive errors:	30 / 93.75 [b]	12 / 80.0	2 / 20.0
Myopic	24 / 75.0	12 / 80.0	2 / 20.0
Hyperopic	6 / 25.0	-	
Visual acuity:			
Perception of light	7 / 21.9	3 / 20.0	2 / 20.0
0.01 - 0.09	13 / 40.6	7 / 46.7	6 / 60.0
0.1 - 0.25	12 / 37.5	4 / 26.7	2 / 20.0
0.3 or better	-	1 / 6.6	-

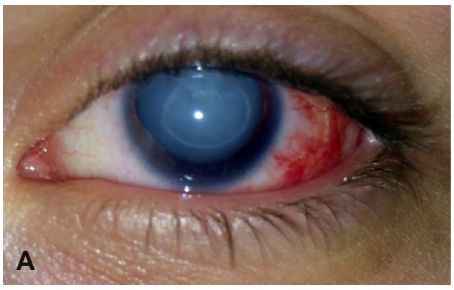
Note: Data are presented as numbers of patients, eyes (n) and percentage. Categorical data are presented as frequencies (n) and percentage (%). Chi-square test was used for analysis of categorical data. p, P-value for differences between groups; [a], children under 11 years and adolescents aged 11 to 18 years; [b], children under 11 years and adults (parents); [c], adolescents aged 11 to 18 years and adults (parents)

majority of cases was subcompensated or decompensated under treatment with hypotensive medications.

Congenital macular hypoplasia (Fig. 8) was observed in 93.75% and 93.3% of the eyes of children and adolescents, respectively, which was confirmed by OCT.

Nystagmus (an involuntary to-and-fro movement of the eyes) was found in most cases (87.5%, 86.7%, and 100%, respectively) in the three groups, and its intensity increased during slit-lamp examination (Table 1).

Refractive errors were found in almost all children and adolescents (93.7% and 80.0%, respectively). Most children and adolescents with refractive errors (75.0% and 80.0%, respectively) were myopic, and 25% of children with refractive errors (these children were of a young age) were hyperopic. In adults, the refraction and fundus assessment was challenging due to central corneal opacity and abnormal transparency of the media (Table 1).



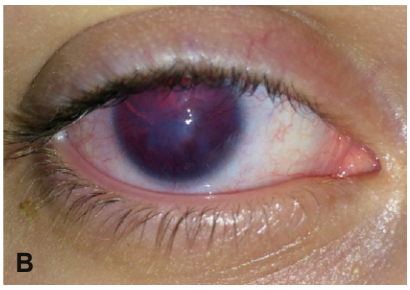


Fig. 3. (A) Stage B aniridia-associated keratopathy and progressive semiresolved subluxated cataract in a 15-year-old adolescent. (B): Stage C aniridia-associated keratopathy involving the central cornea in a 15-year-old adolescent.

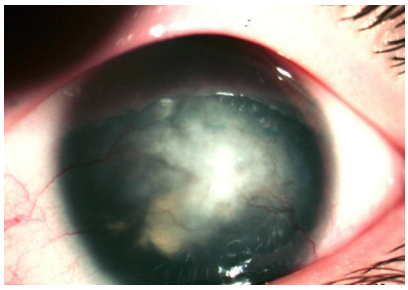


Fig. 4. Stage D aniridia-associated keratopathy in an adult patient of 38 years. Note a large central corneal opacity with corneal conjunctivalization and vascularization.

In groups 1, 2 and 3, visual acuity (VA) was ≤ 0.1 in 40.6%, 46.7%, and 60.0% of eyes, respectively, and 0.1-0.25 in 37.5%, 26.7%, and 20.0% of eyes, respectively. A 12-year-old adolescent had a VA of 0.3 in the eye with partial aniridia and no corneal changes (Fig. 9).

Other congenital ocular abnormalities (anterior microphthalmia, megalocornea, and persistent pupillary membrane [PPM]) were found in isolated cases in all groups (Table 1).

The percentage of familial CA increased from 75% to 100% with increasing age, and sporadic CA was diagnosed only in children (Tables 2, 3). Complete aniridia was more common in patients with familial CA in all groups (75.0%, 86.7%, and 100.0%, respectively), and was found in 62.5% of patients with sporadic CA. Only stage B AAK was found in patients with sporadic CA (25.0%). In familail CA, the severity of corneal changes increased with increasing age. In adolescents with familial CA, stage B AAK was the most common (40.0%), followed by stages C and D AAK (20.0% and 13.3%, respectively); in adults with familial CA, stage C AAK - representing corneal changes with the involvement of the central cornea and apparent corneal vascularization - was the most common (70.0%), followed by stage D AAK (30.0%). Of note, among congenital aniridia eyes of children, congenital cataract was found in all cases of sporadic aniridia and 62.5% of cases of familial aniridia. Progressive cataract was found in 86.7% of the eyes of adolescent patients with familial aniridia, with 5 (33.3%) of these eyes showing lens subluxation. Of the

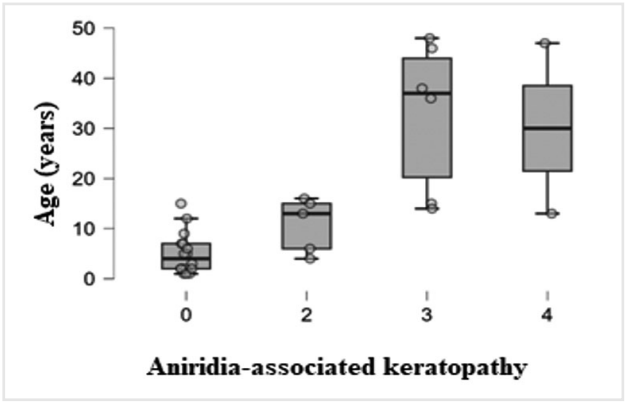
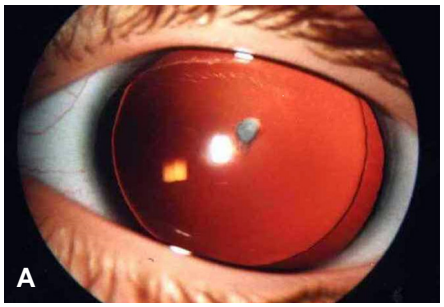
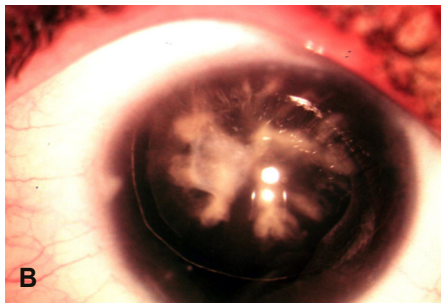


Fig. 5. Box plots showing the presence of aniridia-associated keratopathy in patients of different ages (0, absent; 1, stage A (no patients with stage A in this sample); 2, stage B; 3, stage C; 4, stage D)





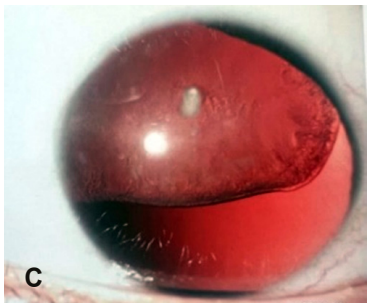


Fig. 6. (A): Congenital complete aniridia with anterior polar cataract in a 24-month infant. (B): Progressive lamellar cataract with a superiorly subluxated lens in an eye with congenital complete aniridia in a 16-year-old adolescent

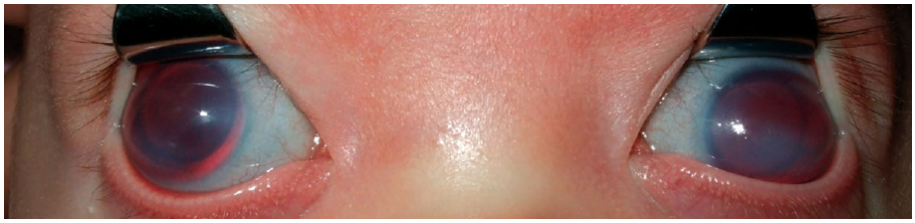


Fig. 7. Congenital glaucoma with corneal edema and buphthalmos in a 7-month infant with congenital aniridia

Table 2. Clinical manifestations of congenital familial aniridia in patients of the study groups (21 patients/ 41 eyes)

Clinical manifestations	Children under 11 years	Adolescents aged 11 to 18 years	Adults (parents)
	8 children (16 eyes) eyes / percentage	8 adolescents (15 eyes) eyes / percentage	5 adults (10 eyes) eyes / percentage
	n / %		
Aniridia (on biomiscrosopy):			
complete	12 / 75.0	13 / 86.7	10 / 100.0
incomplete	4 / 25.0	2 / 13.3	-
Aniridia-associated keratopathy:	-	11 / 73.3 [a]	10 / 100.0 [b]
stage B	-	6 / 40.0 [a,c]	-
stage C	-	3 / 20.0 [c]	7 / 70.0 [b]
stage D	-	2 / 13.3 [c]	3 / 30.0
Corneal edema	2 / 12.5	1 / 6.7	-
Persistent pupillary membrane	6 / 37.5 [a,b]	-	-
Cataract:	10 / 62.5	13 / 86.7 [c]	4 / 40.0
Congenital (anterior polar or anterior capsular)	10 / 62.5 [a,c]	-	-
Progressive (semire-solved, lamellar or calcified)	-	13 / 86.7 [a,c]	4 / 40.0 [b]
Superior lens subluxation	2 / 12.5	5 / 33.3	4 / 40.0
Aphakia	-	-	3
Pseudophakia	-	2	3
Glaucoma:			
Congenital	2 / 12.5	-	-
Secondary	-	9 / 60.0 [a]	6 / 60.0 [b]
Foveolar hypoplasia	14 / 87.5 [a]	14 / 93.3 [c]	-
Optic nerve hypoplasia and partial optic atrophy	2 / 12.5 [a]	10 / 66.7	6 / 60.0 [b]
Nystagmus	16 / 100.0	13 / 86.7	10 / 100.0
Refractive errors			
Myopic	14 / 87.5	12 / 80.0 [c]	2 /20.0 [b]
Hyperopic	-	-	-
Visual acuity:			
Perception of light	3 / 18.8	3 / 20.0	2 / 20.0
0.01- 0.09	8 / 50.0	7 / 46.7	6 / 60.0
0.1-0.25	5 / 31.2	4 / 26.7	2 / 20.0
0.3 or better	-	1 / 6.6	-

Note: Data are presented as numbers of patients, eyes (n) and percentage. Chi-square test was used for analysis of categorical data. p, P-value for differences between groups; [a], children under 11 years and adolescents aged 11 to 18 years; [b], children under 11 years and adults (parents); [c], adolescents aged 11 to 18 years and adults (parents)

eyes of adults with familial aniridia, 40% showed progressive lamellar cataract with lens subluxation, and others had a history of cataract surgery. Signs of congenital glaucoma were equally common (12.5%) in children with sporadic aniridia and those with familial aniridia. Among adolescents and adults, secondary glaucoma was more common in familial aniridia (60.0%) ($p < 0.05$). Foveolar hypoplasia was found in all (100%) children with sporadic CA, and in 87.5% and 93.3%, respectively, of children and adolescents with familial CA (fundus assessment in adults was challenging due to apparent corneal changes). Partial optic atrophy was less common in sporadic CA (37.5%) than in adolescents and adults with familial CA (66.7% and 60.0%, respectively). Nystagmus was the most common symptom in CA, and was found in 75.0% of cases with sporadic CA, and in 100.0%, 86.7%, and 100.0%, respectively, of cases with familial CA in age groups 1 to 3. Refractive errors were also common, and were found in all (100%) aniridic eyes of patients with sporadic CA (62.5% were myopic and 37.5% hyperopic). In addition,

Table 3. Clinical manifestations of sporadic aniridia (8 children/ 16 eyes)

Clinical manifestations	Children under 11 years
	n / %
Aniridia (on biomicrosopy)	
complete	10 / 62.5
incomplete	6 / 37.5
Aniridia-associated keratopathy:	4 / 25.0
stage B	4 / 25.0
stage C	-
stage D	-
Corneal edema	2 / 12.5
Persistent pupillary membrane	4 / 25.0
Cataract:	
Congenital (anterior polar or anterior capsular)	16 / 100.0
Progressive (semiresolved, lamellar or calcified)	-
Superior lens subluxation	-
Aphakia	
Pseudophakia	-
Glaucoma:	2 / 12.5
Congenital	2 / 12.5
Secondary	-
Foveolar hypoplasia	16 / 100.0
Optic nerve hypoplasia and optic atrophy	6 / 37.5
Nystagmus	12 / 75.0
Refractive errors	16 / 100.0
Myopic	10 / 62.5
Hypermyopic	6 / 37.5
Visual acuity:	
Perception of light	3 / 18.8
0.01- 0.09	5 / 31.2
0.1-0.25	8 / 50.0
Better than 0.3	-

Note: Data are presented as numbers of eyes (n) and percentage

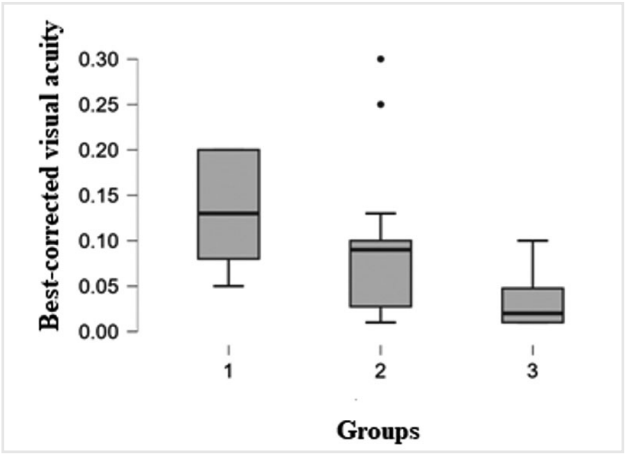


Fig. 9. Visual acuity distributions in age groups. Note: 1, children under 11 years; 2, adolescents aged 11 to 18 years; 3, adults (parents)

myopia was found in 87.5% and 80.0%, respectively, of aniridic eyes of children and adolescents with familial CA. Apparent changes in the anterior and posterior segments resulted in severely reduced VA (< 0.1) in 50.0%, 46.7%, and 60.0%, respectively, in aniridic eyes in patients with familial CA in groups 1 to 3, whereas 50% of patients with sporadic CA had VA ranging from 0.1 to 0.25.

Discussion

The absence of the iris is a classic manifestation of a congenital abnormality of the eye and the first symptom that attracts the attention of the parents, pedia-tricians and ophthalmologists. Aniridia-related iris defects may vary from a rudimentary iris tissue defect to partial aniridia, asymmetric colobomata and sectoral iris atrophy. Such atypical cases of CA require genetic confirmation to clarify the diagnosis and establish the functional prognosis.

In the current study, complete familial aniridia was more common than sporadic aniridia (72.4% vs 27.6%), whereas Gronskov et al [11] and Eden et al [12] found the former form to be less common than the latter form.

Clinicians working at the leading edge of aniridia management believe that the major problem of the disease is not the absence of the iris but the associated ocular complications like AAK (95%) [13, 14], congenital and/or complicated cataract (50-85%) [14], glaucoma (45-75%) [12, 15], macular and optic nerve hypoplasia (79.0-86.0%) [16].

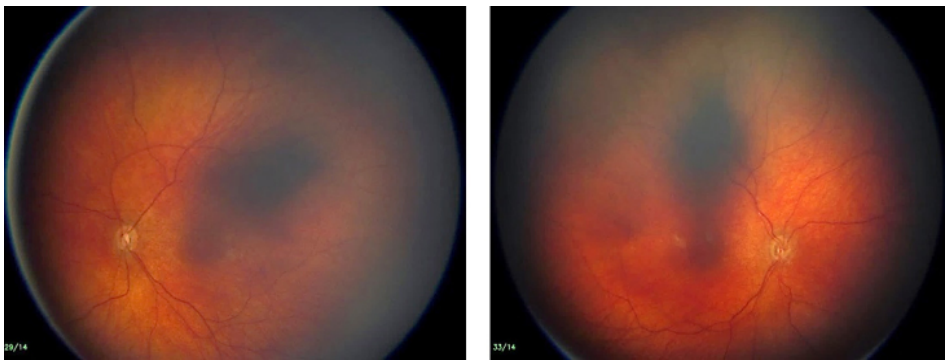


Fig. 8. Macular and optic nerve hypoplasia in a 7-year-old child with congenital aniridia

Our analysis of intrinsic clinical manifestations of CA depending on the child age revealed a number of traits. AAK was very rare and low-grade at an early child age. The age of onset and progression of the first symptoms of AAK can vary and "expressions can begin in the first decade of life" [17]. In a study by Shiple and colleagues [18], the average age of diagnosis of keratopathy in a group of individuals with aniridia was 20 years. Others [12, 19, 20] noted that the youngest individual with aniridia-associated visually disturbing haze was 5 years old. In our group of children under 11 years with CA, the percentage of those showing signs of AAK was relatively small (12.5%). Our youngest patient with AAK (he had sporadic CA and congenital glaucoma) was 4 years old, which is in agreement with the finding by Eden and colleagues [20]. We found that the frequency of AAK tended to increase with age. In our group of adolescents with CA, the percentage of those showing signs of AAK increased to 73.3% (11 eyes), and although stage B AAK was the most common, this group (but not the group of younger patients) included also eyes with stage C or D AAK. In the group of parents of patients, stage C AAK was the most common (70%), followed by stage D (30%).

Congenital cataract is a common manifestation of CA, and the frequency of congenital cataracts in our population of aniridia patients (81.3%) was in agreement with that in a study by Park and colleagues [19] (82.5%) but substantially higher than that found in children and teenagers by Eden and coworkers [12] (53.8%). Congenital cataracts in aniridia eyes may be anterior or posterior polar cataracts or subcapsular cataracts [20]. In the majority of cases, such cataracts are not large and do not cause reduced visual function in the absence of the iris. We found only congenital anterior polar cataracts and anterior capsular cataracts in our aniridia patients. Eden and coworkers [20] observed lens dislocation in 5 of 12 aniridia patients (42%), with the lens displaced upwards in all cases. Singh and colleagues [21] noted subluxation in 55 of 247 (22.3%) phakic eyes with aniridia, and 51 of these 55 lens subluxations (92.7%) were in the superior direction. In the current study, the rate of aniridic eyes with lens subluxation for adolescents of 11 to 18 years was substantially higher than for children under 11 years (33.3% vs 6.25%).

Congenital or secondary glaucoma is a sight threatening manifestation of CA. Congenital glaucoma symptoms usually manifest at infancy whereas secondary glaucoma symptoms usually manifest at adolescence [21]. The development of glaucoma in CA is explained by the presence of apparent goniodygenesis. A significant correlation was found between the magnitude of the iris defect and development of glaucoma [12]. Despite developmental abnormalities of the anterior chamber angle causing glaucoma, congenital glaucoma is rarely seen in patients with CA [12]. In the current study, the rate of congenital glaucoma in eyes of children under 11 years was 12.5%, which is in agreement with that reported by Landsend and colleagues [6] (15%). Adachi and coworkers [23], however, reported that glaucoma evolved in 14 of 29 aniridic eyes (48.2%) before the age of one year. In the current study, the rate of secondary glaucoma in aniridia was 60% for eyes of adolescents as well as eyes of adult parents of child or adolescent patients.

Hypoplasia of the macular fovea is a characteristic ophthalmoscopic finding in aniridia patients, and occurs approximately in 79–92% of subjects [6, 24, 25]. In the present study, the rate of foveolar displasia in aniridia was 93.75% for eyes of child patients under 11 years and 93.3% for eyes of adolescents.

Conclusion

First, CA (iridial hypoplasia) is a rare bilateral panocular genetic disorder, which in 93.3-93.8% of our cases was associated with foveal hypoplasia, resulting in a low visual acuity of light perception to 0.1-0.25. Complications of CA include keratopathy (mostly caused by stem cell deficiency), cataract, glaucoma, and etc.

Second, we found that the most favorable state of the eye was in Ukrainian children aged under 11 years. These children exhibited a relatively low rate of low-grade AAK (12.5%) and an 81.3% rate of congenital cataract, which did not affect visual acuity in the absence of the iris. Additionally, the rate of congenital glaucoma in these children was 12.5%; the disease was accompanied by corneal edema and buphthalmos and was a substantial problem, requiring urgent surgery.

Third, among Ukrainian adolescents aged 11 to 18 years, the state of the aniridian eye was worse than that in children aged under 11 years due to the progression of aniridia-associated complications. Thus, AAK rate increased to 73.3% and severity increased to stage C and even stage D. Additionally, there was progression to lamellar, semi-resolved and even calcified cataracts, which further deteriorated vision, requiring surgery. Of note, among aniridic eyes of adolescents, the rate of lens subluxation was higher than that among aniridic eyes of children aged under 11 years ($p < 0.05$). Secondary glaucoma was of concern, was seen only in adolescents and adults (at a rate of 60%), and was mostly not decompensated or subcompensated despite treatment with medications.

Fourth, parents of aniridic children exhibited the worst state of the eye (however, the study was limited by low numbers of parents and their eyes, 5 and 10, respectively). There were cases exhibiting terminal (D) stage AAK, and some aphakic and peseudophakic eyes had decompensated or subcompensated glaucoma after several surgical procedures.

Fifth, compared to eyes with familial CA, eyes with sporadic CA showed substantially lower rates of complications like AAK (12.5% vs 73.3%, respectively) and glaucoma (12.5% vs 60%, respectively), and these differences were significant ($p < 0.05$).

Lastly, given our findings of progression of major ocular complications of CA with age, early detection of this difficult-to-treat congenital ocular abnormality, consulting for children younger than 12 months under general anes-

thesia at the Pediatric Ophthalmology Department, State Institution "The Filatov Institute of Eye Diseases and Tissue Therapy of the NAMS of Ukraine", and subsequent observation visits every 6 to 12 months depending on the clinical status, are a must. We believe it is warranted to establish a state register of children with CA and develop treatment algorithms and protocols for child patients with CA in order to avoid the severe sequelae which can be seen today in parents of such patients.

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Abbreviation. *AAK, aniridia-associated keratopathy; ACA, anterior chamber angle; CA, congenital aniridia; OCT, optical coherence tomography; PPM, persistent pupillary membrane; the UN, the United Nations; VA, visual acuity*