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Prevalence of glaucoma and characteristics of ocular manifestations in patients with Naevus of Ota

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ABSTRACT

Aims This cross-sectional study aimed to determine the prevalence of glaucoma and associated ocular characteristics in Thai patients with Naevus of Ota, comparing those with ocular melanocytosis or oculodermal melanocytosis to those exhibiting only skin hyperpigmentation.

Methods Patients who were diagnosed with Naevus of Ota at Siriraj Hospital, Thailand, underwent a comprehensive ophthalmic assessment by a glaucoma specialist. Those unable to cooperate in an outpatient setting were examined under general anaesthesia. The assessments comprised visual acuity, intraocular pressure (IOP), anterior segment findings, gonioscopy, corneal diameter (in patients aged <3 years) and fundus examination. Visual field and optical coherence tomography tests were performed as indicated.

Results A total of 163 patients (184 eyes) were examined, including 115 eyes with ocular melanocytosis or oculodermal melanocytosis. The mean age at examination was 15.0±15.6 years. Open-angle glaucoma was identified in 2 eyes (1.1%), ocular hypertension in 6 eyes (3.3%) and glaucoma suspicion in 16 eyes (8.7%). Among those with ocular melanocytosis or oculodermal melanocytosis, ocular hypertension and glaucoma suspicion were more common (4.3% and 12.2%, respectively) than in those with only skin hyperpigmentation (1.4% and 2.9%, respectively).

Conclusion Although the prevalence of glaucoma in Naevus of Ota is low, patients with ocular melanocytosis or oculodermal melanocytosis are at greater risk of ocular hypertension and suspected glaucoma than are those with skin-only hyperpigmentation. Targeted screening, particularly in younger individuals and those requiring examination under general anaesthesia, should be considered. The main limitation of the study is its cross-sectional design, offering only a one-time view of a prolonged clinical progression. Moreover, the use of anaesthetic inhalation could have resulted in lower IOP readings during general anaesthesia.

Trial registration number TCTR20210223004.

WHAT IS ALREADY KNOWN ON THIS TOPIC

⇒ Glaucoma is a significant ocular complication associated with Naevus of Ota, but there is limited evidence regarding the prevalence of glaucoma in individuals with this condition, especially in younger patients.

WHAT THIS STUDY ADDS

⇒ The prevalence of glaucoma was 1.7% among 115 eyes from consecutive Thai patients with ocular melanocytosis or oculodermal melanocytosis. Among the 85 patients who were 10 years old or younger in this study, none had glaucoma and only three had ocular hypertension. Two of these three patients presented with bilateral, extensive cutaneous lesions.

HOW THIS STUDY MIGHT AFFECT RESEARCH, PRACTICE OR POLICY

⇒ The low prevalence of glaucoma must be weighed against the risks and costs of general anaesthesia examinations in children who cannot cooperate in the clinic.

INTRODUCTION

Naevus of Ota, also known as oculodermal melanocytosis (ODM), is a benign melanotic lesion that typically presents from birth. The condition is characterised by a bluish-grey to brown facial patch, typically distributed along the trigeminal nerve branches—especially the ophthalmic and maxillary divisions on the ipsilateral side.[1 2] Hyperpigmentation in Naevus of Ota is attributed to melanocytes trapped in abnormal locations. The condition can extend to various ocular structures, including the episclera, sclera, uvea, cornea, lens, iris, anterior chamber angle, choroid and optic nerve head. Hyperpigmentation can also be found in extraocular sites such as the oral mucosa, meninges, brain and inner ear.[3 4]

Naevus of Ota is particularly prevalent in Asian populations, with reported incidences ranging from 0.014% to 0.034%[5] and a pronounced female predominance (female-to-male ratio of approximately 5:1).[6] Two distinct peaks in onset have been described: the first year of life and the second decade.[6] Its pathophysiology is attributed to embryological failure in melanoblastic cell migration.[7]

The key complications of Naevus of Ota are glaucoma and melanoma formation.[8–10] Glaucoma in Naevus of Ota is often attributed to obstructed aqueous outflow pathways caused by melanocyte accumulation and increased pigmentation within the anterior chamber angle, potentially elevating intraocular pressure (IOP).[10] A specific ocular finding, iris mammillation (small, smooth, uniformly distributed nodules along the pupil margin), is also linked to raised IOP and an increased risk of intraocular melanoma.[11 12] Early and regular ophthalmic assessments for patients with Naevus of Ota have therefore been recommended to mitigate the risk of glaucoma and its accompanying damage.[8 13]

Our study aimed to determine the prevalence of glaucoma and its ocular manifestations in Naevus of Ota. Additionally, we sought to assess the need for early screening, particularly in younger patients, to prevent irreversible visual impairment.

MATERIALS AND METHODS

Study design

This cross-sectional study was conducted at the Department of Dermatology and the Department of Ophthalmology, Faculty of Medicine Siriraj Hospital, between April 2021 and April 2023. It was not possible for patient and public involvement in this study. Detailed study procedures are provided in the online supplemental protocol file.

Patient selection

Patients with a clinical diagnosis of Naevus of Ota who visited the Department of Dermatology and the Department of Ophthalmology, Faculty of Medicine Siriraj Hospital, during June 2005 and June 2023 were listed. From all the patients who were listed, we contacted and scheduled them for an eye examination at least once by a glaucoma consultant (SPe) between April 2021 and April 2023. Demographic data, birthmark characteristics and results of comprehensive ophthalmic assessments were recorded.

Ophthalmic examinations

A single glaucoma consultant conducted all the eye examinations. Each patient's level of cooperation determined the need for general anaesthesia (GA) for the eye examination.

For young or uncooperative patients

In patients requiring GA (eg, concurrent skin laser treatment), the following examinations were performed:

- Visual acuity assessment via the fix-and-follow method or the near vision test.
- Anterior segment evaluation using a handheld slit-lamp microscope.
- Corneal diameter measurement with Castroviejo callipers (if ≤3 years of age).
- IOP measurement with a Perkins tonometer.
- Gonioscopy using a Koppe lens.
- Dilated fundus examination via indirect ophthalmoscopy.

The GA method used in this study is a routine method in the institute. In children, inhalational induction is used. The medications include Nitric Oxide, Desflurane, Midazolam, Thiopental and Succinylcholine. The IOP was measured first after intubation.

For cooperative older children and adults

For patients able to cooperate in an outpatient setting, the following procedures were conducted:

- Visual acuity measurement via an early treatment diabetic retinopathy study chart.
- Anterior segment examination with a standard slit-lamp microscope.
- IOP measurement by Goldmann applanation tonometry.
- Fundus examination using slit-lamp biomicroscopy.

Additional investigations, including optical coherence tomography and Humphrey visual field (HVF) testing, were undertaken at the discretion of the glaucoma consultant.

Table 1 Demographic characteristics and prevalence of glaucoma, ocular hypertension and suspected glaucoma in patients with Naevus of Ota

Variables	Number (%)
163 patients	
Female	120 (73.6)
Age at diagnosis of Naevus of Ota (years): median (IQR)	7.54 (1.13, 22.34)
Age at initial eye examination (years): median (IQR)	8.13 (1.25, 22.67)
Follow-up time (years): median (IQR)	0.5 (0, 3.4)
Case with bilateral lesion	
ODM	9 (5.5%)
ODM/OM*	7 (4.3%)
OM	3 (1.8%)
OM/skin hyperpigmentation†	1 (0.6%)
Skin hyperpigmentation	1 (0.6%)
184 eyes	
Prevalence of glaucoma	2 (1.1)
Prevalence of OHT	6 (3.3)
Prevalence of glaucoma suspected	15 (8.2)
Maximal IOP (mm Hg, n=173): median (IQR)	16.0 (13.0, 19.0)
Cup to disc ratio (n=157)	0.33±0.11
Number of eyes examined under general anaesthesia	30 (16.3%)

*One side with ODM and another side with OM.
†One side with OM and another side with skin hyperpigmentation.
IOP, intraocular pressure; ODM, oculodermal melanocytosis; OHT, ocular hypertension; OM, ocular melanocytosis.

Table 2 Ocular findings in patients with ocular melanocytosis or oculodermal melanocytosis

Ocular findings	Number (%)
Scleral/episcleral involvement	100 (54.3)
Iris hyperpigmentation	26 (14.1)
Iris mammillation	5 (2.7)
Pigmentation at fundus	8 (4.3)

Diagnostic definitions

- *Ocular melanocytosis (OM)*: this condition is characterised by scleral or episcleral melanosis, iris mammillation, trabecular meshwork hyperpigmentation (without identifiable cause), iris hyperpigmentation or fundus pigmentation.[14–16]
- *ODM*: this term describes the combination of OM with ipsilateral deep dermal melanosis of the eyelids, periocular skin or both.[9 16]
- *Hyperpigmentation only*: this term describes skin hyperpigmentation without ocular involvement.
- *Glaucoma*: this condition involves glaucomatous structural damage.[17 18]
  - Category 1: optic nerve head changes with a cup-to-disc ratio ≥0.7, asymmetrical cup-to-disc ratio ≥0.2 between the two eyes, with neuroretinal rim thinning or retinal nerve fibre layer defects with or without corresponding functional loss (visual field defects).
  - Category 2: optic nerve head changes with a cup-to-disc ratio ≥0.9, asymmetrical cup-to-disc ratio ≥0.3 between the two eyes.[15]
  - Category 3: IOP >26 mm Hg with visual acuity less than 3/60 or with previous glaucoma surgery when the optic disc cannot be examined.

For children <18 years of age, the Childhood Glaucoma Research Network (CGRN) will be used following these criteria (two from the listed items).[19]

- IOP >21 mm Hg.
- Cup to disc asymmetry ≥0.2.
- Focal thinning of the optic disc rim.
- Presence of Haab striae, corneal oedema or increased corneal diameter.
- Visual field defect.
- Axial myopia.

In our study, gonioscopy was used to classify glaucoma as open-angle or closed-angle. Other secondary causes were also considered.

- Glaucoma suspect:
  - Eyes with suspicious optic nerve heads (cup-to-disc ratio ≥0.7 but <0.9 or inter-eye asymmetry ≥0.2) with no disc haemorrhage, no rim notching, no retinal nerve fibre layer defect, no definite visual field defect.
  - Abnormal angle findings.
  - Unexplained visual defects suggestive of glaucoma.
  - Unexplained optical coherence tomography suggestive of glaucoma.
  - IOP ≥21 mm Hg without abnormalities in the optic disc, retinal nerve fibre layer or visual field which is defined as ocular hypertension (OHT).[20]

For children <18 years of age, CGRN will be used following these criteria (one from the listed item).[19]

- IOP >21 mm Hg.
- Cup to disc asymmetry ≥0.2.
- Focal thinning of optic disc rim.
- Presence of Haab striae, corneal oedema or increased corneal diameter.
- Visual field defect.

Statistical analysis

Categorical data are presented as frequencies and percentages. Continuous variables that were not normally distributed are presented as medians and IQRs. The Mann-Whitney U test was used to compare IOP values between eyes with and without OM. The statistical analyses were performed with PASW Statistics, V.18 (SPSS, Chicago, Illinois, USA).

RESULTS

Demographic and clinical characteristics

A summary of the demographic data is presented in table 1. Most patients were female (73.6%) and had unilateral involvement (87.8%). Approximately 52.5% of patients were younger than 10 years, and 39% were younger than 5 years at their initial examination. Among the 184 eyes studied, 115 exhibited OM or ODM, whereas the remaining eyes presented only skin hyperpigmentation. Table 2 summarised ocular findings in patient with melanocytosis or ODM.

Table 3 Prevalence of glaucoma, OHT, and glaucoma suspects in eyes with hyperpigmentation only and OM or ODM

	Diagnosis: eyes (%)					
	Normal	Suspected	OHT	Glaucoma	Total	P value
Hyperpigmentation only	66 (95.7)	2 (2.9)	1 (1.4)	0	69 (100.0)	0.057
OM or ODM	94 (81.7)	14 (12.2)	5 (4.3)	2 (1.7)	115 (100.0)	
Total	160 (87.0)	16 (8.7)	6 (3.3)	2 (1.1)	184 (100.0)	

ODM, oculodermal melanocytosis; OHT, ocular hypertension; OM, ocular melanocytosis.

Table 4 Detailed ocular findings in patients diagnosed with glaucoma or ocular hypertension and patients suspected of having glaucoma

Patient no.	Age range (years)/sex	IOP max	Conjunctival melanosis	Hyperpigmented iris	Iris mammillation	Hyperpigmentation via gonioscopy	C:D	Pigmented fundus	Diagnosis
1	40–45/F	15	No	Yes	Yes	Yes	0.5	No	NTG
2	12–18/F	9	Yes	No	No	Yes	0.6	No	Secondary glaucoma
3	<1/F	22	Yes	No	No	Yes	0.2	No	OHT
4	6–11/F	21	Yes	No	No	Yes	0.1	No	OHT
5	6–11/F	27	Yes	No	No	Yes	0.1	No	OHT
6	6–11/M	23	Yes	No	No	No	0.3	No	OHT
7	6–11/M	22	Yes	No	No	No	0.3	No	OHT
8	45–50/F	14	Yes	No	No	No	0.7	Yes	Glaucoma suspect (enlarge cupping)
9	6–11/F	17	No	No	No	Yes	0.4	No	Glaucoma suspect (asymmetrical cupping)
10	55–60/M	18	Yes	Yes	No	No	0.4	No	Glaucoma suspect (occludable angle)
11	12–18/F	24	No	No	No	Yes	0.5	No	Glaucoma suspect (suspicious OCT)
12	12–18/F	20	Yes	Yes	No	Yes	0.7	No	Glaucoma suspect (enlarge cupping)
13	20–25/F	19	Yes	No	No	Yes	0.5	No	Glaucoma suspect (asymmetrical cupping)
14	20–25/F	19	Yes	Yes	No	Yes	0.5	No	Glaucoma suspect (suspicious OCT)
15	20–25/F	17	Yes	Yes	No	Yes	0.5	No	Glaucoma suspect (asymmetrical cupping)
16	20–25/F	15	Yes	No	No	No	0.6	No	Glaucoma suspect (asymmetrical cupping)
17	30–35/F	18	Yes	Yes	Yes	Yes	0.5	No	Glaucoma suspect (asymmetrical cupping)
18	40–45/F	17	No	No	No	Yes	0.6	No	Glaucoma suspect (asymmetrical cupping)
19	40–45/F	15	No	No	No	Yes	0.6	No	Glaucoma suspect (asymmetrical cupping)
20	40–45/F	11	Yes	Yes	No	No	0.6	No	Glaucoma suspect (asymmetrical cupping)
21	12–18/M	19	No	Yes	No	No	0.6	Yes	Glaucoma suspect (asymmetrical cupping)

C:D, cup-to-disc ratio; F, female; IOP, intraocular pressure; M, male; NTG, normal-tension glaucoma; OD, right eye; OHT, ocular hypertension; OS, left eye.

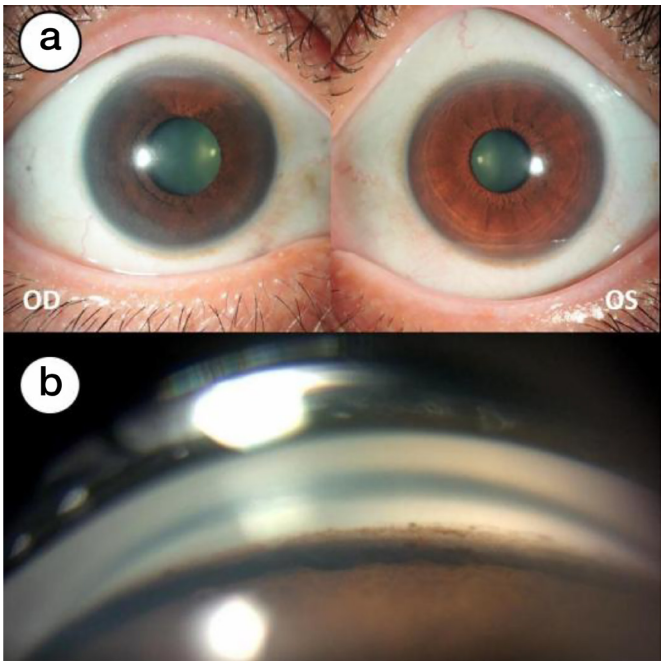

Figure 1 Anterior segment and gonioscopy findings in a patient with normal tension glaucoma. (a) Anterior segment photograph of the right eye showing iris hyperpigmentation and mammillations. Iris hyperpigmentation and mammillations were observed in the right eye. (b) Gonioscopy image depicting prominent trabecular meshwork hyperpigmentation.

Prevalence of glaucoma, OHT and glaucoma suspects

The prevalence rates of glaucoma, OHT and glaucoma suspects are shown in table 3. In OM and ODM patients, among the 14 eyes classified as glaucoma suspects, 2 displayed enlarged cupping, 9 had asymmetrical cupping, 2 had suspicious OCT and 1 presented narrow angles.

IOP comparisons

The mean maximal IOP was 16.0 mm Hg (range, 13.0–19.0 mm Hg). Eyes with ODM had a significantly greater maximal IOP than those without oculodermal melanocytosis. The median (IQR) values were 17.0 (13.0–19.0 mm Hg) mm Hg and 15.0 (12.8–18.0) mm Hg, respectively (p=0.013).

Ocular hyperpigmentation and the glaucoma spectrum

Eyes were divided into two groups on the basis of the presence of ocular hyperpigmentation (table 3). Among eyes with OM and ODM, 18.2% were diagnosed with glaucoma or suspected of having glaucoma or OHT. In contrast, 4.3% of the eyes with only skin hyperpigmentation were suspected of having glaucoma or were diagnosed with OHT, and none of these eyes had glaucoma. The mean age at the initial examination of this subgroup was 24.3±16.8 years (range, 9.9 months–41.7 years). Table 4 details the ocular findings of the eyes that were diagnosed with glaucoma or OHT and those that were suspected of having glaucoma.

Visual field testing

Visual field was tested in 46 eyes. There was an unremarkable visual field in 45 eyes and an abnormal visual field in 1 eye (patient no. 16 in table 4). The 24-2 visual field has 1 point of central field defect; however, it was confirmed unremarkable in HVF 10-2.

All patients with glaucoma and glaucoma suspects had been tested for visual field, except for four patients, three were children and one was lost to follow-up.

Case presentations

Patient 1: normal-tension glaucoma

The first example patient is a 43-year-old woman who was diagnosed with normal-tension glaucoma in the right eye. She exhibited OM manifesting as iris hyperpigmentation and iris mammillation without scleral involvement (figure 1a). Gonioscopy revealed grade 4 angles in both eyes (modified Shaffer's system) with prominent trabecular meshwork hyperpigmentation on the right side (figure 1b). The left eye was normal. The diagnosis of normal-tension glaucoma was based on glaucomatous optic neuropathy and a relatively elevated IOP of 15 mm Hg in the affected eye compared with 8 mm Hg in the contralateral eye. HVF testing (HVF 24-4) was unremarkable. Optical coherence tomography revealed a superior retinal nerve fibre layer defect in the right eye.

Patient 2: secondary glaucoma and OHT

The second example patient is a 12-year-old girl with secondary glaucoma in the left eye and OHT in the right eye. She had undergone left trabeculectomy at another hospital before presentation. Examination revealed skin hyperpigmentation and conjunctival melanocytosis in the left eye but without iris hyperpigmentation. The vertical cup-to-disc ratios were 0.4 in the right eye and 0.6 in the left eye. The IOPs were 23 mm Hg (right) and 9 mm Hg (left). Gonioscopy revealed grade 4 angles in both eyes, with marked trabecular meshwork hyperpigmentation in the left eye only. The right eye displayed no such hyperpigmented features.

DISCUSSION

In this study, the prevalence of glaucoma was 1.7% among 115 eyes from consecutive Thai patients with OM or ODM. We focused on younger age groups with OM or ODM, and the median age at the initial eye examination was 8.13 years (IQR 1.25–22.67). Notably, 39% of the patients were 5 years old or younger.

The combined prevalence of glaucoma and OHT was 6.0%. Compared with a previous study, which reported a 10.3% prevalence of glaucoma or OHT in ODM patients,[21] our rate was lower. One possible explanation involves changing treatment trends for Naevus of Ota. Modern laser therapies for pigmented skin lesions encourage parents to seek medical care for their children at younger ages, thus reducing the average age at diagnosis. In the past, when no effective treatment existed,

patients often sought attention only after developing ocular concerns.

Several factors may explain the low prevalence of glaucoma in younger patients. In addition to the reasons already discussed, pathogenesis itself could provide insight. Glaucoma is multifactorial, and its progression often requires time before clinical manifestation. Alterations in the extracellular matrix within the trabecular meshwork can increase aqueous outflow resistance, eventually contributing to elevated IOP and glaucoma development.[22] Age may also play a significant role, as strong evidence indicates that the incidence of glaucoma increases with advancing age.[23] Although the pathogenesis of glaucoma in Naevus of Ota remains poorly understood, some studies suggest that pigment deposition in the trabecular meshwork may impede aqueous outflow. Others argue that Naevus of Ota alone is not sufficient to induce glaucoma.[10 24] Further long-term follow-up studies are needed to determine the incidence of glaucoma in older OM and ODM cohorts, as well as to elucidate specific risk factors and pathogenic mechanisms.

Evaluating young children for glaucoma can be challenging. Detailed examinations often require GA, which entails additional time, costs and health risks.[25–27] Among the 85 patients who were 10 years old or younger in this study, none had glaucoma and only three had OHT. Two of these three patients presented with bilateral, extensive cutaneous lesions. Although glaucoma screening cannot be abandoned entirely in young children with OM or ODM, clinicians might tailor their approach. More frequent examinations may be warranted in patients with extensive bilateral lesions, as these individuals may carry a greater risk of glaucoma. A new ocular classification system has been proposed, which presents an intriguing potential as a tool for assessing the risk of glaucoma.[28] Nevertheless, the identification of a glaucoma risk factor was not feasible in this study due to the limited sample size and the low prevalence of glaucoma within the cohort. Another notable finding is that all glaucoma patients presented IOP-dependent characteristics. One patient had OHT in the right eye and glaucoma in the left eye with a functioning filtering bleb. Another patient, who was diagnosed with normotensive glaucoma in the right eye, had an IOP of 15 mm Hg in the affected eye compared with 8 mm Hg in the unaffected eye. These observations align with previous studies showing that patients with Naevus of Ota can experience prolonged elevated IOP.[10 24 29] In clinical practice, simple IOP measurement could be an initial screening tool for patients with Naevus of Ota. However, the findings of this study demonstrate a low median maximum IOP. Potential contributing factors include: first, the low prevalence of glaucoma and OHT; second, the examination of 30 eyes under GA, where the GA methodology employed may have led to falsely low IOP readings; and finally, the presence of a functioning bleb in glaucomatous cases, which may contribute to reduced IOP.

Pigmentation may also contribute to glaucoma pathogenesis, especially when it extends into the eye. This hypothesis is supported by the absence of glaucoma in patients who presented with only skin hyperpigmentation. Furthermore, the combined prevalence of OHT and suspected glaucoma in patients with OM or ODM was 4.3% and 12.2%, respectively, whereas it was 1.4% and 2.9%, respectively, in patients who had only cutaneous hyperpigmentation. According to a population-based survey of Thais over 50 years of age, the prevalence rates of OHT and glaucoma suspects were 2.14% and 4.14%, respectively.[24] Although this older population differs from our study cohort, the higher OHT and glaucoma suspect rates associated with OM and ODM remain noteworthy.

A key limitation of this study is its cross-sectional design, which provides only a single snapshot of a potentially prolonged clinical course. More robust data may emerge from future longitudinal studies. Additionally, anaesthetic inhalation may have lowered IOP measurements obtained under GA. To mitigate this issue, we relied on careful optic nerve head evaluation rather than solely on IOP values. Nevertheless, caution is necessary when interpreting the average IOP and OHT prevalence derived from measurements collected under anaesthesia.

Another factor is our recruitment approach. We primarily enrolled patients scheduled for laser therapy in the dermatology department, and these individuals typically lacked ocular symptoms. In contrast, public announcements and direct recruitment from ophthalmology clinics might preferentially attract patients already experiencing eye problems.

Finally, none of our patients exhibited systemic syndromes such as phakomatosis pigmentovascularis or Klippel-Trenaunay-Weber syndrome. Several reports confirm that these conditions significantly increase the risk of glaucoma.[30–35] These syndromes may represent severe forms of phakomatoses involving components such as port-wine stains and haemangiomas combined with ocular hyperpigmentation.[32 34 36–39]

Future screening strategies for young patients with OM or ODM should be tailored. The low prevalence of glaucoma must be weighed against the risks and costs of GA examinations in children who cannot cooperate in the clinic. Larger, longitudinal studies are needed to inform future screening recommendations.

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Competing interests None declared.

Patient and public involvement Patients and/or the public were not involved in the design, or conduct, or reporting, or dissemination plans of this research.

Patient consent for publication Consent obtained directly from patient(s).

Ethics approval This study involves human participants. The protocol received Siriraj Institutional Review Board approval (SIRB protocol number 1038/2563(IRB4), COA no.Si 037/2021) and complied with the tenets of the Declaration of Helsinki. Informed consent was obtained prior to each patient examination.

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REFERENCES

1 Fitzpatrick TB, Kitamura H, Kukita A, *et al*. Ocular and dermal melanocytosis. *AMA Arch Ophthalmol* 1956;56:830–2.
2 Sinha S, Cohen PJ, Schwartz RA. Nevus of Ota in children. *Cutis* 2008;82:25–9.
3 Swann PG, Kwong E. The naevus of Ota. *Clin Exp Optom* 2010;93:264–7.
4 Teekhasaenee C, Ritch R, Rutnin U, *et al*. Ocular findings in oculodermal melanocytosis. *Arch Ophthalmol* 1990;108:1114–20.
5 Chan HHL, Kono T. Nevus of Ota: clinical aspects and management. *Skinmed* 2003;2:89–96.
6 Hidano A, Kajima H, Ikeda S, *et al*. Natural history of nevus of Ota. *Arch Dermatol* 1967;95:187–95.
7 Agarwal P, Patel BC. Nevus of ota and ito. In: *StatPearls*. Treasure Island (FL): StatPearls Publishing LLC, 2024.
8 Plateroti AM, Scavella V, Abdolrahimzadeh B, *et al*. An Update on Oculodermal Melanocytosis and Rare Associated Conditions. *Semin Ophthalmol* 2017;32:524–8.
9 Abdolrahimzadeh S, Pugi DM, Manni P, *et al*. An update on ophthalmological perspectives in oculodermal melanocytosis (Nevus of Ota). *Graefes Arch Clin Exp Ophthalmol* 2023;261:291–301.
10 Magarasevic L, Abazi Z. Unilateral open-angle glaucoma associated with the ipsilateral nevus of ota. *Case Rep Ophthalmol Med* 2013;2013:924937.
11 Ragge NK, Acheson J, Murphree AL. Iris mammillations: significance and associations. *Eye (Lond)* 1996;10 (Pt 1):86–91.
12 Sánchez Marugán B, Acebes García M, García Hinojosa J, *et al*. Iris mammillations. Three case reports. *Archivos de La Sociedad Española de Oftalmología (English Edition)* 2014;89:279–81.
13 Rujimethapass N, Manuskiatti W, Wanitphakdeedecha R, *et al*. Ocular manifestations of facial port-wine stain, nevus of Ota, and phakomatosis pigmentovascularis in Asian patients. *J Am Acad Dermatol* 2021;85:1194–200.
14 Ota M, Tamino H. A variety of nevus frequently encountered in Japan, nevus fusco-coeruleus ophthalmomaxillaris and its relation to pigmentary changes in the eye. *Tokyo Med J* 1939;63:1242–4.
15 Fitzpatrick TB, Zeller R, Kukita A, *et al*. Ocular and Dermal Melanocytosis. *Arch Ophthal* 1956;56:830–2.
16 Carreño E, Saornil MA, Garcia-Alvarez C, *et al*. Prevalence of ocular and oculodermal melanocytosis in Spanish population with uveal melanoma. *Eye (Lond)* 2012;26:159–62.
17 Foster PJ, Buhrmann R, Quigley HA, *et al*. The definition and classification of glaucoma in prevalence surveys. *Br J Ophthalmol* 2002;86:238–42.
18 Mannaf SMA, Islam MS, Islam MN, *et al*. Population-based survey of the prevalence and types of glaucoma in Bangladesh. *BMJ Open Ophth* 2024;9:e001609.
19 Freedman S, Ely A. Childhood glaucoma. In: *Handbook of pediatric retinal OCT and the eye-brain connection*. Elsevier, 2020: 313–34.
20 Kass MA, Heuer DK, Higginbotham EJ, *et al*. The Ocular Hypertension Treatment Study: a randomized trial determines that topical ocular hypotensive medication delays or prevents the onset of primary open-angle glaucoma. *Arch Ophthalmol* 2002;120:701–13.
21 Teekhasaenee C, Ritch R, Rutnin U, *et al*. Glaucoma in oculodermal melanocytosis. *Ophthalmology* 1990;97:562–70.
22 Miyazaki M, Segawa K, Urakawa Y. Age-related changes in the trabecular meshwork of the normal human eye. *Jpn J Ophthalmol* 1987;31:558–69.
23 Zhang N, Wang J, Li Y, *et al*. Prevalence of primary open angle glaucoma in the last 20 years: a meta-analysis and systematic review. *Sci Rep* 2021;11:13762.
24 Foulks GN, Shields MB. Glaucoma in oculodermal melanocytosis. *Ann Ophthalmol* 1977;9:1299–304.
25 Wang X, Xu Z, Miao CH. Current Clinical Evidence on the Effect of General Anesthesia on Neurodevelopment in Children: An Updated Systematic Review with Meta-Regression. *PLoS ONE* 2014;9:e85760.
26 Robinson EJ, Lyne TC, Blaise BJ. Safety of general anaesthetics on the developing brain: are we there yet? *BJA Open* 2022;2:100012.
27 Jáuregui-Huerta F, Redolar-Ripoll D, Cupul-García JC, *et al*. General anesthesia in infants: neurobiological and neuropsychological concerns. *Bol Med Hosp Infant Mex* 2020;77:54–67.
28 Vishnevskia-Dai V, Moroz I, Davidy T, *et al*. Naevus of Ota: clinical characteristics and proposal for a new ocular classification and grading system. *Br J Ophthalmol* 2021;105:42–7.
29 Pal E, Chaine G, Hershkovitch D, *et al*. Nevus of Ota associated with chronic glaucoma. *J Fr Ophtalmol* 1997;20:771–4.
30 Wu H-E, Li T-N, Jin C-L, *et al*. Neurofibromatosis Type 1 with Becker's Nevus and Nevus of Ota. *Indian J Dermatol* 2020;65:81–3.
31 Namiki T, Takahashi M, Nojima K, *et al*. Phakomatosis pigmentovascularis type IIb: A case with Klippel-Trenáunay syndrome and extensive dermal melanocytosis as nevus of Ota, nevus of Ito and ectopic Mongolian spots. *J Dermatol* 2017;44:e32–3.
32 Yang Y, Guo X, Xu J, *et al*. Phakomatosis Pigmentovascularis Associated With Sturge–Weber Syndrome, Ota Nevus, and Congenital Glaucoma. *Medicine (Baltimore)* 2015;94:e1025.
33 Abdolrahimzadeh S, Plateroti AM, Recupero SM, *et al*. An Update on the Ophthalmologic Features in the Phakomatoses. *J Ophthalmol* 2016;2016:3043026.
34 Petchyim S. A Congenital Glaucoma Associated with Phakomatosis Pigmentovascularis in Infant Case Report. *Siriraj Med J* 2017;69:217–9.
35 Abdolrahimzadeh S, Pugi DM, de Paula A, *et al*. Ocular manifestations in phakomatosis pigmentovascularis: Current concepts on pathogenesis, diagnosis, and management. *Surv Ophthalmol* 2021;66:482–92.
36 Abdolrahimzadeh S, Scavella V, Felli L, *et al*. Ophthalmic Alterations in the Sturge-Weber Syndrome, Klippel-Trenaunay Syndrome, and the Phakomatosis Pigmentovascularis: An Independent Group of Conditions? *Biomed Res Int* 2015;2015:786519.
37 Patil B, Sinha G, Nayak B, *et al*. Bilateral Sturge-Weber and Phakomatosis Pigmentovascularis with Glaucoma, an Overlap Syndrome. *Case Rep Ophthalmol Med* 2015;2015:106932.
38 Teekhasaenee C, Ritch R. Glaucoma in phakomatosis pigmentovascularis. *Ophthalmology* 1997;104:150–7.
39 Abdolrahimzadeh S, Fameli V, Mollo R, *et al*. Rare Diseases Leading to Childhood Glaucoma: Epidemiology, Pathophysiogenesis, and Management. *Biomed Res Int* 2015;2015:1–11.