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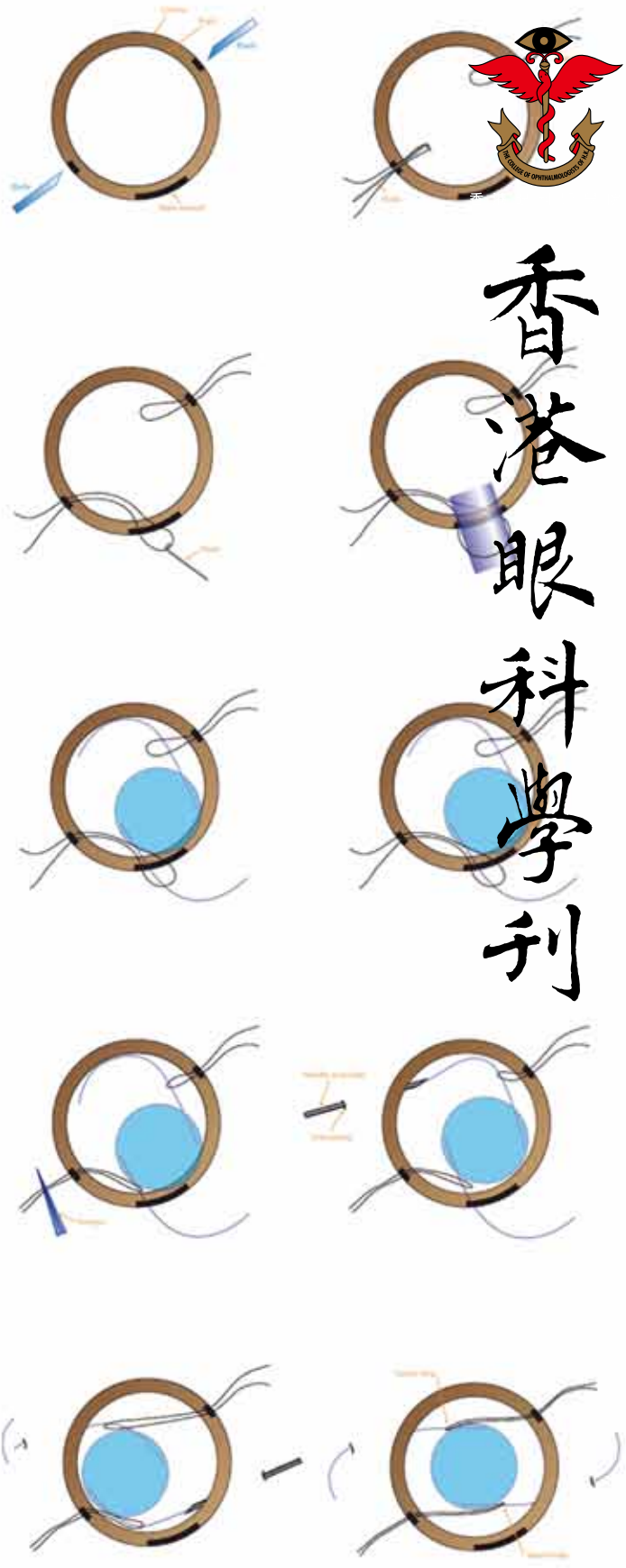

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Results from a 6-month, controlled, contribution-of-elements study (n=560). Patients with open-angle glaucoma (including pseudoexfoliation or pigment dispersion component) and/or ocular hypertension with elevated IOP were given SIMBRINZA Suspension (brinzolamide 1%/brimonidine tartrate 0.2% ophthalmic suspension) BID or brinzolamide ophthalmic suspension, 1% BID; or brimonidine tartrate ophthalmic soution, 0.2% BID. Patients' baseline mean diurnal IOP was 26mm Hg. At baseline SIMBRINZA Suspension n=193; brinzolamide n=191; brimonidine n=175.

Reference: 1. Katz G, DuBiner H, Samples J, et al. Three-month randomized trial of fixed-combination brinzolamide, 1%, and brimonidine, 0.2%. JAMA Ophthalmol. 2013;131(6):724-30 **2.** Hong Kong Product Insert **3.** Aung T, Laganovska G, Hernandez Paredes TJ, et al. Twice-Daily Brinzolamide/Brimonidine Fixed Combination versus Brinzolamide or Brimonidine in Open-Angle Glaucoma or Ocular Hypertension. Ophalmology. 2014:121(12):2348-55.

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1. *International Committee of Medical Journal Editors (ICMJE). Uniform requirements for manuscripts submitted to biomedical journals: writing and editing for biomedical publication. ICMJE; April 2010. www.icmje.org*

香 港 眼 科 醫 學 院

THE HONG KONG JOURNAL OF OPHTHALMOLOGY
BEST ORIGINAL ARTICLE AWARD

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To encourage submissions to the *Hong Kong Journal of Ophthalmology* (HKJO) by trainees and fellows of the College of Ophthalmologists of Hong Kong (COHK).

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Two awards will be given out every year. Each awardee will be given a $1000 cash prize (HK dollars), which is supported generously by the Timothy KC Liu Fund.

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- Trainees refer to those who have not been conferred the title 'FCOphthHK' at the time of submission.
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- Original articles (including, but not limited to, prospective or retrospective clinical studies, observational studies, epidemiological studies, basic science studies, meta-analysis, etc) published in the HKJO during the year will automatically enter the selection process.
- Case reports (with case number <3), review articles and letters to the editor will not be eligible for this award.
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- Scientific merit
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EDITORIAL

香 港 眼 科 醫 學 院

Antithrombotic treatment in ophthalmic surgery

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Ophthalmic surgery is generally performed in elderly people, many of whom are on systemic medications such as antiplatelet and anticoagulant therapies. These drugs are routinely discontinued preoperatively to prevent severe intra- and post-operative bleeding. Nonetheless, premature discontinuation of antiplatelet therapy or interruption of anticoagulation increases the risk of thromboembolic complications including death. It is important to balance the risk of thrombotic complications and the benefit of avoiding intra- and post-operative bleeding.

Antiplatelet drugs such as aspirin and clopidogrel are prescribed for patients with atherosclerotic vascular disease and/or for prevention of acute coronary syndrome, myocardial infarction, or stroke. Patients with coronary artery disease are often prescribed dual antiplatelet therapy that combines aspirin and clopidogrel for a mandated duration after an acute coronary syndrome or stent (drug-eluting or bare-metal) implantation. Premature discontinuation of antiplatelet therapy may lead to major cardiac complications and thrombotic events (myocardial infarction, stent thrombosis, and non-fatal stroke) and even death.[1]

Anticoagulant treatments such as vitamin K antagonist (e.g. warfarin) and the novel oral anticoagulants (NOACs) [e.g. dabigatran, rivaroxaban, apixaban, and edoxaban] have been widely used in patients with atrial fibrillation to prevent stroke or systemic embolism, in patients with valvular heart disease after mechanical prosthetic heart valve replacement, and for venous thromboembolism prophylaxis. Patients on anticoagulants are risk-stratified for better perioperative anticoagulant management. The risk of thromboembolic events in patients with a history of atrial fibrillation can be assessed using the CHADS2 score. The thrombotic risk is highest in those with mitral valve prosthesis, older aortic valve prosthesis, and recent stroke or transient ischemic attacks or venous thromboembolic events. As interruption of anticoagulation therapy increases the risk of potentially fatal thromboembolic events, the medical team needs to consider the thrombotic risk based on patient-related and surgery-related factors before continuing or stopping anticoagulation. Oral anticoagulants are generally continued perioperatively for minor procedures, whereas invasive procedures may require discontinuation of oral anticoagulants or bridging anticoagulation therapy with subcutaneous low-molecular-weight heparin or intravenous unfractionated heparin during interruption of anticoagulation.

Cataract surgery is commonly performed in elderly patients on antithrombotic therapy. The management of antiplatelet and anticoagulation therapies perioperatively in cataract surgery has been well-studied.[2-4] Cataract removal is an avascular procedure with topical anesthesia, with or without

intracameral anesthesia. No significantly increased risk of severe bleeding or sight-threatening bleeding complications is associated with antithrombotic treatment, although there may be an increase in the incidence of non-significant or self-limiting hemorrhage such as hyphema or subconjunctival hemorrhage. A Hong Kong study reported no difference in hemorrhagic complications between cases continuing or withholding NOAC during phacoemulsification.[5]

Vitreoretinal (VR) surgery also appears to be safe in patients on antithrombotic therapy. Nonetheless, the literature lacks evidence with strong statistical power to formulate guidelines on perioperative management of antithrombotic therapy for VR surgeries. Anticoagulant drugs are associated with an increased risk of intraocular hemorrhage, but not with serious consequences, re-operations, or surgery failure.[6-8] In a systematic review and meta-analysis, NOACs were reported to reduce the risk of intraocular bleeding by approximately 20% when compared with warfarin.[9] Further studies of the effects of NOACs on the outcome of VR surgery are needed. In patients on warfarin, VR surgery has been shown to result in no anesthesia-related or intraoperative hemorrhagic complications despite some postoperative hemorrhages that resolved spontaneously. Similarly, in systemically anticoagulated patients receiving intravitreal injections, the risk of hemorrhagic complications is extremely low; cessation of anticoagulation therapy is not needed.[10-12] The benefits of continuing antithrombotic therapy outweigh the risks of discontinuation.

Trabeculectomy or a glaucoma drainage device may result in large variation in intraocular pressure during and after glaucoma surgery. Anticoagulation can thus increase the risk of certain vision-threatening complications such as suprachoroidal hemorrhage. Bleeding can negatively impact the surgical outcome and lead to surgical failure. Intraoperative bleeding, particularly in the subconjunctival space, may impede surgeon visualization of tissues. The proinflammatory nature of bleeding can result in excessive scarring. Blood clots can mechanically block the drainage site at the sclerostomy or tube opening. Patients on anticoagulation therapy have been reported to have an increased postoperative hyphema rate, with or without a significant impact on the final surgical outcome.[13,14] As the evidence for altering anticoagulation therapy for glaucoma surgery is lacking, different surgeons adopt different practices. Consultation with the primary prescriber of anticoagulation therapy is recommended before alteration.

There is a lack of evidence on which to base guidelines for perioperative management of oculoplastic patients on antithrombotic therapy. The incidence of serious hemorrhagic

Table 1. Perioperative management for antiplatelets

Antiplatelet	Timing of last dose before surgery	Postoperative resumption
Aspirin	5 days	Next day
Clopidogrel (non-coronary stent)	10 days	Next day
Clopidogrel (coronary stent)	- Continue if within 6 weeks (bare stent) or 12 months (drug-eluting stent) of stent implantation - Consult physicians if need to be stopped	
Dural antiplatelets (non-coronary stent)	Continue aspirin and stop clopidogrel 10 days before surgery	
Dual antiplatelets (coronary stent)	- Avoid surgery within 6 weeks (bare stent) or 12 months (drug-eluting stent) of stent implantation - Consult physician if need to be stopped	
Others (e.g. dipyridamole)	Consult physicians	

Table 2. Perioperative management for warfarin

Risk of thrombo-embolism	Management	Bridging therapy	Postoperation
Low	- Omit warfarin 5 days before surgery - INR test 1-2 days before surgery	Not applicable	- Resume warfarin 12-24 hours after surgery
Moderate	No bridging		
	Consider bridging: - Omit warfarin 5 days before surgery - Daily INR test after warfarin stopped; start bridging when INR <1.5	Prophylactic dose: - Enoxaparin 40 mg daily subcutaneous injection; half dose 1 day before surgery	- Resume therapeutic dose enoxaparin and warfarin according to bleeding risk* - Daily INR test; stop enoxaparin when INR >1.5
High	Need bridging: - Omit warfarin 5 days before surgery - Daily INR test after warfarin stopped; start bridging when INR <1.5	Therapeutic dose: - Enoxaparin 1.5 mg/kg daily subcutaneous injection; half dose 1 day before surgery	

Abbreviation: INR = international normalized ratio

** Low risk: resume therapeutic dose of enoxaparin after 24 hours; high risk: delay therapeutic dose of enoxaparin 48 to 72 hours after surgery when there is adequate hemostasis*

Table 3. Risk stratum for thromboembolism

Risk of thrombo-embolism	Mechanical heart valve	AF	VTE
Low	Bileaflet aortic prosthesis without AF and no other risk factors for stroke	CHADS2 score* of 0 to 2 (and no prior stroke or TIA)	Single VTE occurred >12 months ago and no other risk factors
Moderate	Bileaflet aortic prosthesis without AF and one of the following: AF, prior stoke or TIA, hypertension, diabetes, congestive HF, age >75 years	CHADS2 score* of 3 to 4	- VTE within past 3 to 12 months - Non-severe thrombophilic conditions - Recurrent VTE - Active cancer (treated within 6 months or palliative)
High	- Any mitral valve prosthesis - Older aortic valve prosthesis - Recent (within 6 months) stroke or TIA - Pregnancy with mechanical heart valve	- CHADS2 score* of 5 or 6 - Recent (within 3 months) stroke or TIA - Rheumatic valvular heart disease	- Recent (within 3 months) VTE - Severe thrombophilia

Abbreviations: AF = atrial fibrillation, HF = heart failure, TIA = transient ischemic attack, VTE = venous thromboembolism
** CHADS2 score: congestive heart failure (1 point), hypertension (1 point), age >75 years (1 point), diabetes (1 point), and prior stroke or transient ischemic attack (2 points)*

Table 4. Perioperative management for patients on novel oral anticoagulants

Novel oral anticoagulant	Timing of last dose before surgery	
	Standard bleeding risk	High bleeding risk
Creatinine clearance (mL/min)*		
Dabigatran		
>50	24 hours	2 days
>30 to ≤50	2 days	4 days
≤30	4 days	6 days
Rivaroxaban		
>30	24 hours	2 days
<30	2 days	4 days
Postoperative resumption		
Dabigatran (150 mg twice daily)†	Resume 24 hours after surgery	Resume 48-72 hours after surgery
Rivaroxaban (20 mg daily)†	Resume 24 hours after surgery	Resume 48-72 hours after surgery

** (140 – age in years) x mass (kg) x constant (1.23 for men; 1.04 for women) / serum creatinine (μmol/L)*
† Reduced dose for impaired renal function; for patients at high risk of thromboembolism, consider starting anticoagulant at reduced dose on evening after surgery or the day after

complications secondary to oculoplastic procedures is low.[15] Patients on antithrombotic agents or not are comparable in terms of perioperative hemorrhagic complications.[16] In patients undergoing oculoplastic procedures with a greater hemorrhagic risk such as dacryocystorhinostomy or orbital decompression, surgeons generally suggest stopping antithrombotic agents if it is safe to do so.[17] Alteration of the antithrombotic therapy should be discussed with the patient and individualized based on the nature of surgery and the patient's medical condition and need for anticoagulation.

For most types of ophthalmic procedures, the risks of interrupting antithrombotic therapy appear to outweigh the benefits. Aspirin does not appear to be associated with an increased risk of significant bleeding in patients undergoing any type of ophthalmic surgery. Similarly, warfarin does not appear to increase the risk of bleeding in ophthalmic surgery when the international normalized ratio is within the normal range. In glaucoma or oculoplastic surgeries where bleeding at the operative site may impact the surgical outcome, perioperative alteration of anticoagulation therapy may be performed cautiously and should involve the opinion of a medical specialist. Nonetheless, the evidence on which to base precise recommendations about NOACs is lacking. Further investigations are needed to explore the risk of significant bleeding when altering NOAC therapy.

A collaboration between the Hong Kong Eye Hospital and the Department of Medicine, Queen Elizabeth Hospital aims to design a guideline to standardize and simplify the perioperative antithrombotic management in ophthalmic surgeries. The suggested algorithms for perioperative management of antiplatelet, warfarin, and NOAC therapies are shown in **Tables 1** to **4**.[18-21]

Declaration

The authors have no conflicts of interest to disclose.

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REVIEW ARTICLE

Idiopathic intracranial hypertension: adult versus pediatric patients

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Abstract

Idiopathic intracranial hypertension (IIH), also known as benign intracranial hypertension or pseudotumor cerebri, is characterized by increased intracranial pressure secondary to unknown causes. IIH affects both adults and children but predominately young obese females of child-bearing age. IIH gives rise to headache, visual disturbances, and symptoms of raised intracranial pressure. The pathophysiology of IIH remains elusive; postulations include obesity-related hormonal changes, obstruction of the central venous sinus, and hereditary causes. Diagnosing IIH is based on modified Dandy criteria. Diagnosis in children is challenging as symptoms and presentations vary; atypical and delayed presentations are not uncommon. Treatment for IIH includes oral acetazolamide or lumbar puncture; surgical treatment with shunts and optic nerve sheath fenestration is reserved for complicated and recurring cases. Treatment options for children are limited and associated with considerable risks and side effects. Vision loss is debilitating and occurs in 10% of IIH patients. A thorough and collaborative approach in managing IIH is needed. This study reviews the epidemiology, clinical presentation, diagnosis, and management of IIH in adults and pediatric patients.

Key words: Headache; Idiopathic intracranial hypertension with papilledema; Obesity; Pseudotumor cerebri

Epidemiology

Idiopathic intracranial hypertension (IIH) can occur at all stages of life irrespective of sex but has a predilection towards obese females.[1] The annual incidence of IIH is 0.9 cases per 100,000 people and 3.5 cases per 100,000 obese females of child-bearing age.[2] The incidence of IIH in Asians is lower. In a Japanese cohort, the incidence of IIH was 0.3 cases per 100,000 people per year.[3] The incidence in Chinese is not well studied but is believed to be infrequent.[4] Observational studies have shown a direct relationship between increased body mass index and the risk of IIH.[5] The incidence is expected to increase with the global obesity epidemic. Patients with recent weight gain of 5% to 15% have a higher risk of developing IIH than those with stable weight.[5] About 10% of IIH patients are men who are usually diagnosed a decade older than women.[6] Black patients seem to have a worse vision prognosis than whites. It is unknown whether Asians have the same risks of developing IIH as other races.[7,8]

Childhood IIH occurs most frequently after puberty.[9] Pre-pubertal IIH is uncommon and is rare in neonates and preschool children. The estimated incidence of childhood IIH is 0.8 cases per 100,000 children. In Hong Kong, the incidence was 0.78 case per 100,000.[10] These figures are probably underestimated because of selection bias and diagnostic problems. Post-pubertal IIH patients are mostly obese females, whereas pre-pubertal IIH patients are usually non-obese, with males and females equally affected.[11,12] A retrospective multicenter study of pediatric IIH using anthropometric parameters identified 3 subgroups of patients: young children (girls aged <7 years and boys aged <8.5 years) who were not overweight; early adolescents (boys and girls aged 7-12 years) who were taller and overweight; and late adolescents (boys and girls aged ≥12.5 years) who were overweight.[13] It is unclear whether these 3 subgroups represent natural progression of pediatric IIH or distinct subgroups of disease with different pathophysiology. The association of adiposity and increased linear growth acceleration in post-pubertal, early and late adolescent

IIH patients echoes that of adult IIH cohorts. Contributing factors other than adiposity have yet to be determined. Nevertheless, the overall trend in childhood obesity and metabolic syndromes may increase the incidence of pediatric IIH.

Pathophysiology

The association between obesity and IIH is indisputable, although its exact mechanism remains poorly understood. Observational and epidemiological studies have described a possible association of increased intracranial pressure (ICP) with abnormal vitamin A metabolism. Theory related to hypervitaminosis A and IIH is conflicting. It is speculated that raised vitamin A level in cerebrospinal fluid (CSF) reduces brain water absorption by increasing all trans-retinoic acid gene expression at arachnoid granulation cap cell, ependymal and glial cells.[14] Although earlier reports revealed raised retinol level in CSF of IIH patients, the recent Idiopathic Intracranial Hypertension Treatment Trial (IIHTT) reported that CSF vitamin A toxicity was an unlikely contributing factor.[15-17] The research focus of IIH has shifted to consideration of adipose tissue as an actively secreting endocrine organ.[18] Adipose tissue–derived retinol-binding protein and cytokines like leptin are elevated in obese individuals.[19,20] It is unclear whether the presence of such metabolites has a role in IIH development, but their identification may provide information about disease pathology.

Intracranial obstructions relating to impaired venous drainage is another pathophysiology of IIH. Observational studies have displayed distal narrowing and stenosis of the transverse cerebral sinus in most IIH cases.[21-24] Venous sinus stenosis may induce a cycle of venous outflow obstruction, venous hypertension, reduced CSF absorption, and subsequent raised ICP and further compression to the venous system.[1] It is not known whether this anatomical finding is incidental, causal, or secondary to raised ICP. Transverse venous stenosis is also found in some patients without IIH and may have no functional significance.[25] Regardless of the cause, endovascular stenting at the site of transverse venous stenosis has shown to reduce venous congestion, improve CSF absorption, and reduce ICP.[26] It remains debatable whether the presence of transverse sinus stenosis affects the natural history and prognosis of IIH.

Certain medications (such as oral contraceptive pills, tetracycline, danazol, tamoxifen, lithium, cyclosporin) and a history of polycystic ovary syndrome have been proposed to cause IIH but the evidence is limited owing to small sample sizes.[27-29] Other factor includes CSF homeostasis involving mutation of the aquaporin-4 channel that controls brain water movement via transmembrane osmotic shift.[30] Magnetic resonance spectrometry with metabolic profiling of CSF in IIH has been used to observe pathophysiological mechanisms.[31] There is still no unifying pathophysiological explanation, and further studies are required.

Clinical features

Symptoms of raised ICP are commonly present in IIH. Headache is documented in 62% to 91% of patients and is characterized by frontal, severe, pulsatile pain that is worse when lying supine or aggravated by Valsalva maneuver during early morning.[32-34] Often mistaken as migraine, IIH can also be confused with tension headache and analgesic-related headache, except that IIH headache is continuous. In a large prospective cohort study, headache was the most common presentation (89%).[35] This can be further complicated when IIH headache is superimposed on headaches of other origin. Reduction of Headache Impact Test – 6 score corresponds to disability related to IIH headache.[36,37] Visual abnormalities are often the sole complaint in IIH. Visual field defect is present in 80% to 86% of patients.[35,38,39] Blind spot enlargement is the most frequent symptom (80%), followed by peripheral arcuate defects (72%) and constrictions (54%), associated with nerve fiber type damage and depressed perimetric mean deviation visual field sensitivity (-7.7 dB).[39] This finding is echoed by the IIHTT.[35] Visual acuity loss is usually minimal and is unaffected early in the course. Studies have reported diminished visual acuity (logMAR ≥0.2) in 17% of patients and abnormal color vision (<5 out of 6 in Hardy-Rand-Rittler charts) in 14%.[39-41] Permanent visual field loss is uncommon.[42] Transient visual obscurations, characterized by amaurosis lasting for half a minute, occur at a mean of once per month. It is non-specific to IIH and is unrelated to the presenting visual acuity or final visual outcome, although it might represent transient central retinal artery compression.[35] Other ophthalmic symptoms include reduced color vision (thought to be a result of papilledema)[40] and pulsatile tinnitus (probably caused by turbulent flow from the venous stenosis over transverse sinus),[43] which occurs in 60% of IIH patients on alternate days bilaterally.[35] Other symptoms include vomiting, retrobulbar pain, photophobia, radicular tenderness over the neck and shoulders, and diplopia.[35]

IIH is often diagnosed incidentally in the absence of symptoms. Papilledema is a hallmark in the diagnosis of IIH. The degree of optic disc swelling is graded according to the Frisen scale where grade 0 is normal disc, grade 1 is very early papilledema, grade 2 is early papilledema, grade 3 is moderate papilledema, grade 4 is marked papilledema, and grade 5 is severe papilledema.[44] Early papilledema is most frequently seen (33%).[35,39] Typically occurring in both eyes, unilateral papilledema and IIH without papilledema have been well described in adults.[45,46] They are often accompanied by optic disc changes such as hyperemia of the optic disc and absent spontaneous venous pulsations. The etiology of unilateral cases is unknown. Optical coherence tomography has been used to measure the peripapillary average retinal thickness to identify subtle disc swelling.[47] Nonetheless, its role in children has yet to be evaluated, as reference for pediatric parameters is lacking. Relatively low intraocular pressure in the eye has been reported to predispose to retrolaminar hypoperfusion and result in

disc swelling.[40,45] Nonetheless, a case series demonstrated no significant intraocular pressure difference between papilledemic eyes.[39] Papilledema tends to resolve after 3 to 6 months of therapy.[48] Very infrequently IIH patients go on to develop optic atrophy as a result of long-standing papilledema and disease progression, and thus afferent pupillary defect is uncommon (5.4%).[35,39,49] Decreased abduction related to raised ICP may be present.[50] Choroidal-retinal folds and macular edema are rare.[45]

In children the symptoms are generally similar. Patterns in adolescents are akin to those in adults. Young children have different presentations. Headache is often intermittent and episodic rather than continuous.[51] Verbalizing symptoms of headache can be difficult because of non-specificity and misinterpretation as dizziness, neck and shoulder tenderness, or generalized fatigue. Differentiating IIH headache from intracranial neoplasm is by exclusion only. Brain tumor headaches tend to be nocturnal, deep aching, intermittent, and upon rising.[52] Other suspicion of pyramidal neurological deficits should indicate a diagnosis of central etiology. The pattern of field defect is similar and occurs in 91% of pediatric patients.[32,38,53] Nonetheless, expression problems may delay detection of symptoms, and some patients may compensate field loss by head tilt and accommodation. Up to 6% to 20% of children present with visual acuity loss.[33,38,54] Other visual complaints include photophobia and transient vision loss described as 'shimmering lights with colored centers'.[34] The pattern of papilledema is similar to that in adults. Care should be taken to differentiate papilledema from disc swelling secondary to infective causes, ischemic optic neuropathy, demyelination, pseudo-papilledema such as congenital dysplastic disc or Leber hereditary optic neuropathy. Some children with IIH may present without papilledema,[46] with an estimated prevalence of 5.7%.[55,56] In infants <18 months of age, a patent fontanelle or open sutures may have contributed to the absence of papilledema.[54,57] In older pediatric patients, optic nerve sheath abnormalities have been hypothesized. In any case, the absence of papilledema tends to give a better visual outcome.[58] Proptosis secondary to chronic raised ICP has been described in adults but has not been observed in IIH pediatric cohorts.

Diagnostic challenges

The modified Dandy criteria is the gold standard for diagnosing IIH.[59] It comprises 4 key elements: (1) the presence of signs and symptoms of increased ICP (headaches, nausea, vomiting, transient visual obscurations, papilledema), (2) the lack of localizing focal neurological signs except unilateral or bilateral abducens nerve paresis, (3) CSF opening pressure of ≥25 cm H_2O but without cytological or chemical abnormalities, and (4) neuroimaging adequate to exclude cerebral venous thrombosis. When IIH is suspected, clinicians should collect a thorough medical history of the patient, particularly the onset and characteristics of raised ICP symptoms, relevant drug history, followed by detailed documentation of their visual acuity, color vision, fields, ocular motility, pupillary examination, slit lamp and fundal examination, and neurological examination (**Figures 1** and **2**). Visual field testing should also be performed if permissible. Humphrey 30-2 (Swedish interactive thresholding algorithm) is preferred over 24-2 for improved sensitivity.[56] This should be followed by imaging of the head to exclude a secondary intracranial cause, and confirmed with diagnostic lumbar puncture for opening pressure. It is also advisable to perform ocular coherence tomography for the optic disc and retinal nerve fiber layer and repeat it at intervals for serial monitoring.

There are limitations to these diagnostic criteria in adults and children. The symptoms should only reflect those of raised ICP or papilledema. Headache, double vision, and pulsatile tinnitus represent typical symptoms of IIH. Less typical symptoms such as neck, shoulder radicular pain or diplopia should be handled with caution. Non-specific symptoms such as nausea, vomiting, and photophobia may exist. Transient visual obscuration and visual field loss may only be exhibited in the presence of papilledema. When early central visual field loss is encountered without signs of chronic papilledema, other causes such as inflammatory optic neuropathy should be considered. When seizure, focal neurological complaints, or encephalopathy are involved, central etiologies should be suspected.

The diagnosis of IIH can be established even if the patient is asymptomatic but exhibiting relevant signs. Atypical signs include third nerve palsy, intranuclear ophthalmoplegia, squint, and meningism.[59] These should indicate the diagnosis of intracranial neoplasm, subacute meningoencephalitis, central sinus thrombosis, or venous infarction other than IIH.[60] There are reports describing diagnosis of IIH without signs of papilledema. Caution should also be taken for possible anatomical variance of optic nerve head vasculature, optic nerve sheath abnormality, and pre-existing glaucomatous optic neuropathy.

Accurate measurement of raised ICP could be challenging in patients with IIH. Diagnosis of IIH requires a lumbar opening pressure of ≥25 cm H_2O with the patient lying in the lateral decubitus position with legs extended to eliminate Valsalva maneuver.[61] Typically the mean CSF pressure in IIH is raised at 35 cm H_2O and has no sex or weight predilection.[56] ICP of 20-25 cm H_2O is considered undiagnostic.[62] Lumbar puncture should be repeated in highly suspicious cases owing to the dynamic nature of ICP.[63]

Adequately excluding secondary causes of intracranial hypertension is important. Computer tomography remains the first line of investigation to exclude hydrocephalus and space-occupying lesions. Magnetic resonance imaging (MRI) with contrast may be superior in the detection of specific IIH changes and to exclude leptomeningeal infiltration and isodense tumors. Emerging evidence has shown that IIH exhibits specific radiological signs: (1) flattening of the posterior pole of the eyes, (2) dilatation and tortuosity of the

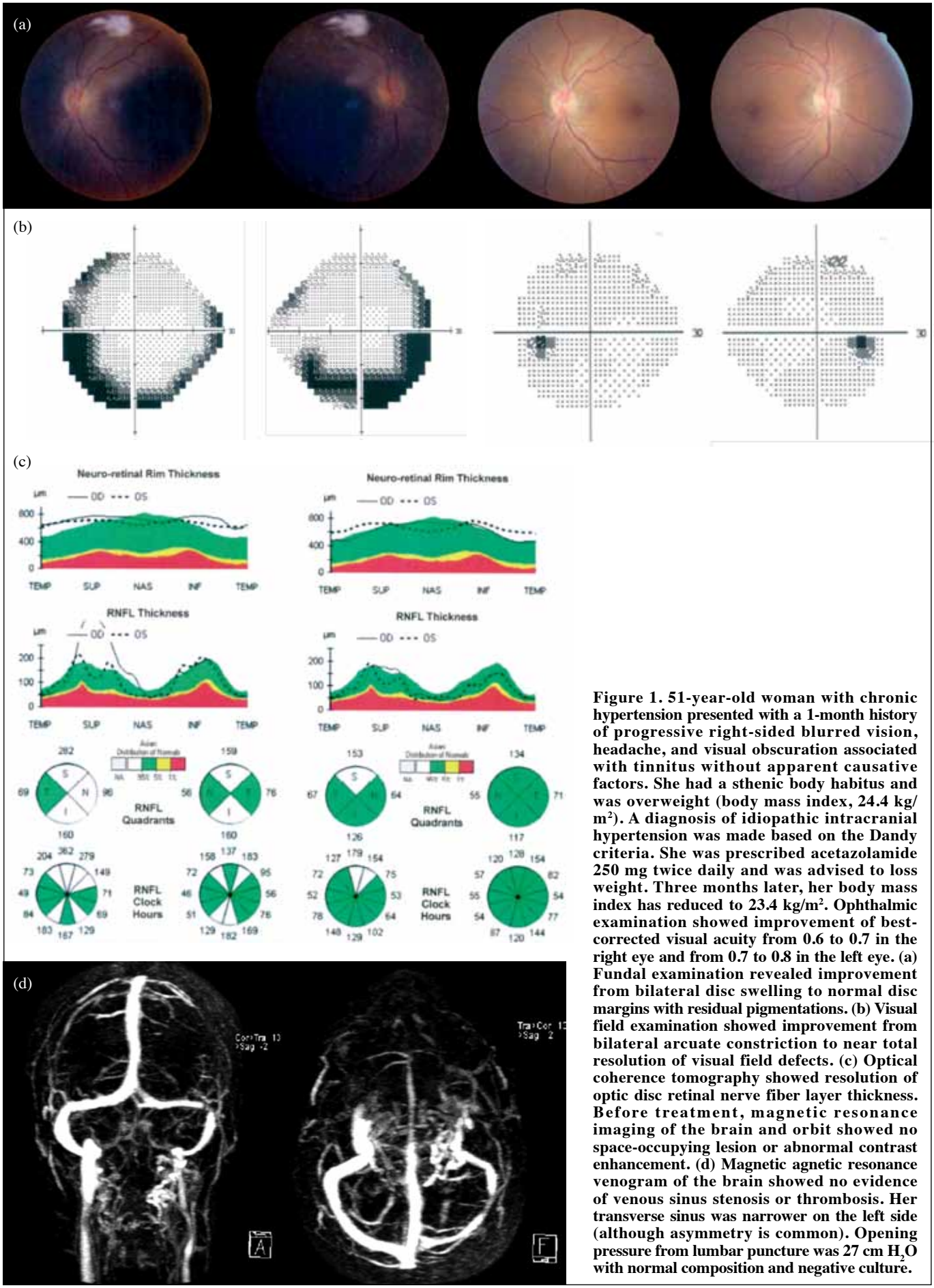

Figure 1. 51-year-old woman with chronic hypertension presented with a 1-month history of progressive right-sided blurred vision, headache, and visual obscuration associated with tinnitus without apparent causative factors. She had a sthenic body habitus and was overweight (body mass index, 24.4 kg/m^2). A diagnosis of idiopathic intracranial hypertension was made based on the Dandy criteria. She was prescribed acetazolamide 250 mg twice daily and was advised to loss weight. Three months later, her body mass index has reduced to 23.4 kg/m^2. Ophthalmic examination showed improvement of best-corrected visual acuity from 0.6 to 0.7 in the right eye and from 0.7 to 0.8 in the left eye. (a) Fundal examination revealed improvement from bilateral disc swelling to normal disc margins with residual pigmentations. (b) Visual field examination showed improvement from bilateral arcuate constriction to near total resolution of visual field defects. (c) Optical coherence tomography showed resolution of optic disc retinal nerve fiber layer thickness. Before treatment, magnetic resonance imaging of the brain and orbit showed no space-occupying lesion or abnormal contrast enhancement. (d) Magnetic agnetic resonance venogram of the brain showed no evidence of venous sinus stenosis or thrombosis. Her transverse sinus was narrower on the left side (although asymmetry is common). Opening pressure from lumbar puncture was 27 cm H_2O with normal composition and negative culture.

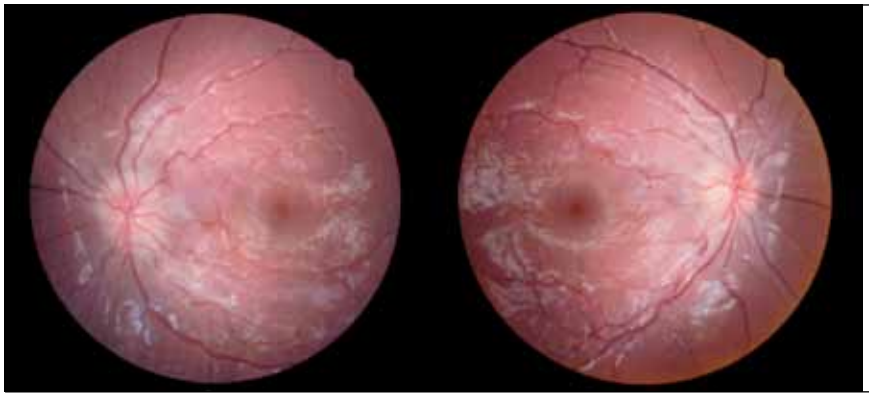

Figure 2. Russel Silver syndrome mimicking idiopathic intracranial hypertension in a 5-year-old boy who had an incidental finding of bilateral disc swelling. He was developmentally delayed, short-statured and failed to thrive. Brain imaging and lumbar puncture opening pressure were normal. He was asymptomatic and had normal visual field. He was suspected to have Russel Silver syndrome (a genetic condition affecting chromosomes 7 and 11), which is characterized by short stature, triangular face, clinodactyly, hemi-hypertrophy, hypoglycemia, and failure to thrive.

optic nerve sheaths, (3) empty sella turcica, and (4) stenosis of one or both transverse cerebral venous sinuses.[1,64] When interpreting images, it is important to differentiate central venous sinus thromboses from transverse venous stenosis, which is more specific to IIH. There is no consensus on routine screening with MRI. When uncertain, MRI venogram should also be used. In pediatric patients, MRI is the choice for diagnosing IIH, although issues with sedation, contrast allergy, renal impairment, and presence of an implanted medical device may preclude its use. In addition, secondary causes of raised ICP should be ruled out, such as Addison's disease, obstructive sleep apnea, pulmonary hypertension, and medications such as contraceptives. MRI should also be used in 'atypical' patients: males, non-obese, elderly, or pre-pubertal children.[65-67]

Diagnosis in children is also based on the modified Dandy criteria, but application of adult-derived criteria to all age groups is disputable. Children present a different spectrum of diagnostic challenge owing to non-specific complaints and variability in symptoms, and they may articulate insufficiently. This may explain why children have more visual damage because of delayed diagnosis. Pediatric patients may exhibit atypical signs such as cerebellar and long-track signs that mimic a posterior fossa space-occupying lesion. This may be further complicated by behavioral changes such as head banging and generalized non-specific irritability or solemnness.[46]

The norm and cut-off opening ICP that satisfies the diagnosis of IIH in children are difficult to determine. ICP varies with age: 8.2 cm H2O represents a normal opening pressure in infants, slowly increasing to 18 cm H_2O in children aged 8 years, eventually reaching an upper limit of 20 cm H_2O in children >8 years old.[68,69] In 2010, a diagnostic ICP for IIH was proposed: an opening pressure of >20 cm H_2O in children <8 years old and >25 cm H_2O in those >8 years old.[55] Nonetheless, a false high opening pressure may be observed in crying children or anxious patients. Lumbar puncture under light sedation is recommended. Fluctuations in ICP should be taken into account and a child may not meet the diagnostic ICP.[63,70,71] In cases of a high suspicion of IIH, a second opening pressure may be necessary. In rare circumstances a lumbar drain with a transducer has been used for monitoring.[72]

The onset of symptoms should also be considered. Patients with rapid onset or fulminant progressive deterioration should raise a concern for cerebral vasculopathy. Nevertheless, conditions that masquerade as IIH should be ruled out. Conditions previously thought to be associated with IIH such as medications (oral contraceptive pills, retinoids), lupus, and vasculitis are now believed to be related to venous sinus thrombosis rather than IIH as a result of hypercoagulability.[73,74] If in doubt, ocular retinal nerve fiber layer scanning may pick up subtle changes in the optic disc.[47,75] Serial fundus photographs and fluorescent angiogram can also be used.

Treatment

The goals of managing IIH are vision preservation and alleviation of symptoms. Weight loss remains crucial as the first-line and long-term management and its effects linger. The trend of weight control is an indicator for recurrence of disease. A weight loss of 5% to 10% results in control of symptomology, signs, and ICP.[76] Other weight-control methods such as dietetic strategies (low salt, low calorie diet), exercise regimens, and bariatric surgery could be considered.[1,77] Treatment of endocrine and metabolic morbidities such as obesity and diabetes may have dual effects of weight reduction and theoretic alteration of homeostasis of ICP. Lumbar puncture is indicated for diagnosis and treatment, and is usually the first attempt for symptom relief.[78] This mode of therapy can have a lasting effect for a few months to years, with some patients requiring only one treatment. Patients with persistent symptoms and signs usually undergo serial lumbar punctures.

In patients whose visual function is impaired, medication such as acetazolamide can be prescribed in parallel. Acetazolamide, a carbonic anhydrase inhibitor, can reduce cerebral fluid production. Combination therapy with a carbonic anhydrase inhibitor and weight loss is efficacious. The multicenter, double-blind, randomized controlled IIHTT showed that the use of acetazolamide with a low-sodium weight-reduction diet resulted in modest improvement in visual field function and papilledema grade at 6 months compared with diet alone. Nonetheless, the long-term effects and functional correlation of visual field improvement are unknown. The recommended dose

of acetazolamide for adults is 1-2 g/day in divided doses, titrated to a maximum dose of 4 g/day.[79] In pre-pubertal children, the dose is started at 15-25 mg/kg/day and titrated to a maximum dose of 100 mg/kg/day.[58,80-82] Post-pubertal children are generally treated with an adult dose.[83] Dose of acetazolamide can be up-titrated as long as it is required and clinically tolerated. Nonetheless, sole use of acetazolamide achieves varying success, and its efficacy in children remains unknown. Moreover, the side-effects of acetazolamide include hypokalemia, metabolic acidosis, development of renal calculi, transaminitis, pancreatitis, and failure to thrive in long-term use.[48] Mineralocorticoid, in the form of methylprednisolone at 15 mg/kg, is another treatment option for acute vision loss.[84] Intravenous injection at a high dose can be used for severe vision loss and to delay surgical treatment.[85] In children, methylprednisolone can be prescribed at 2 mg/kg/day for 2 weeks and tapered over 2 weeks. Other oral medications such as topiramate, zonisamide, octreotide, mannitol, and frusemide can be used when other medications are contraindicated.[86-88] There is no consensus on treatment duration. It has been suggested that medications be continued until visual function and symptoms have stabilized for at least 6 months.[89] Headache usually resolves in response to lowered ICP. Long-term headache can be treated with breakthrough analgesic such as acetaminophen, non-steroidal anti-inflammatory agents or prophylactic agents such as propranolol, amitriptyline, topiramate, and valproate. It is important to be alert for analgesic headache and rebound headache. Non-steroidal anti-inflammatory agents should be avoided in children for fear of Reye's syndrome.[90] Resistant cases invariably require surgical treatment.

Surgical intervention (optic nerve sheath fenestration [ONSF] and CSF shunting) is recommended in fulminant and intractable cases where vision loss is imminent.[85] ONSF is the treatment of choice in cases with acute deterioration in visual function alone.[91,92] ONSF can be performed medially or laterally through incision of the arachnoid dura via a transconjunctival orbitotomy approach. ONSF may stabilize visual acuity, visual field, reduce papilledema and headaches.[91-93] Bilateral stabilization of visual function after unilateral ONSF has also been reported.[94,95] Adverse effects of ONSF include permanent vision loss, pupillary dysfunction, and recurrence. Up to 12% to 33% of cases demonstrate vision improvement initially but deterioration later.[96,97]

In patients with intractable headache and acute progressive vision loss, CSF drainage using a ventriculo-peritoneal shunt or lumbo-peritoneal shunt is preferred. CSF shunting can halt progression of vision loss and is the most widely performed surgery for IIH.[98-100] Current evidence does not suggest superiority of one procedure over another. Nonetheless, ventriculo-peritoneal shunts have a higher failure rate, whereas lumbo-peritoneal shunts need more revision.[101] In addition, CSF shunting is associated with risks of infection, intracranial bleeding, and acquired Chiari malformation.[98] The long-term outcome of visual function has yet to be determined.

Endovascular stenting also improves symptoms and preserves vision in selected cases of transverse venous stenosis by reducing venous pressure and ICP.[26,102] Placement of the stent over the distal transverse sinus or transverse-sigmoid sinus junction is followed by dual antiplatelet therapy for 6 months, and then aspirin for 1-2 years. Nonetheless, it is associated with serious complications such as in-stent migration, re-stenosis, subdural hemorrhage, and infection.[26,102] Given its risk and morbidity, endovascular stenting should be offered only to cases of profound vision loss, fulminant symptoms where medical treatment has been exhausted and when ONSF and CSF shunting are contraindicated.

Management in pediatric cases is mostly medical. Most pediatric patients respond to initial lumbar puncture at diagnosis.[89] Serial lumbar punctures are often unwelcomed owing to the associated pain and need for admission, sedation, and monitoring.[103] The current first-line treatment with carbonic anhydrase inhibitor is acetazolamide. The problems with long-term carbonic anhydrase inhibitors are electrolyte imbalance, failure to thrive, and renal calcification. All can be further complicated by non-compliance. The treatment duration should be the same as for adults. Substitution with frusemide or topiramate may be needed when acetazolamide is ineffective or contraindicated. Weight control remains the long-term goal as for adult IIH patients.[104] Rarely surgical options are applied in children with severe headache and fulminant visual function deterioration. ONSF and CSF shunting have shown varying success. Problems related to children specifically in lumbo-peritoneal shunting are shunt migration, mechanical fracture, and obstruction due to longitudinal growth of the spine.[98,105] Experience in stenting of a transverse venous stenosis in pediatric patients is lacking. Surgical procedures for pediatric IIH should be limited to selective patients.

Prognosis

Permanent and severe vision loss has been reported in 6% to 9% of adults and 10% of children with IIH.[12,33,53,106,107] The incidence of blindness secondary to IIH is approximately 1% to 2%.[108] The degree of obesity is proportional to the amount of vision loss.[109] Other visual complaints such as transient visual obscurations, papilledema, duration of symptoms, and presence of cranial nerve palsies are not associated with worse visual outcomes.[43,50,110] In a few patients, symptoms of headache persist even after management of raised ICP.[111]

The rate of recurrence of IIH is 8% to 10% in adults and 6% to 22% in children.[12,33,53,71,112,113] Recurrence typically occurs within 2 to 5 years of diagnosis, with no sex predilection. Multiparous women seemed to have more recurrence.[39] Changes in visual acuity and field are usually the chief complaint in recurrence and typically occur in those who lose weight initially and regain thereafter.[109,114] Weight control is predictive of recurrence.[109] A weight gain of 6% is associated with recurrence.[114] Obesity in pre-pubertal children increases the rate of recurrence five-fold.[115] Strict weight management is therefore imperative to minimize

fluctuations. Frequent monitoring of visual function, body mass index, and compliance is indicated.

Conclusion

IIH is an uncommon neuro-ophthalmological entity that may result in permanent visual disability. Recognition of clinical symptoms and the use of imaging modalities are essential in diagnosing IIH and excluding secondary causes. Pediatric cases are rare and diagnosis remains difficult owing to variability of symptoms and non-specificity. Diagnosis in atypical patients such as males and non-obese patients should be made with caution. Treatment options include weight loss, lumbar puncture, and acetazolamide. Surgery is reserved for refractory cases. Most visual acuity and field loss are mild and reversible but depend on weight control, recurrence, and timing of diagnosis. The trend of IIH is expected to increase due to the global endemic of obesity. The management of pediatric and adult cases are different and should be individualized.

Declaration

The authors have no conflicts of interest to disclose.

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ORIGINAL ARTICLE

Prevalence of eyelid diseases among adults in Hong Kong

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Abstract

Introduction: Eyelid diseases affect visual function and quality of life. This is the first study to investigate the prevalence of eyelid diseases among the general population in Hong Kong.

Methods: On 22 October 2017, The College of Ophthalmologists of Hong Kong and The Hong Kong Ophthalmological Society jointly organized a Public Education Day. A total of 165 participants were recruited for eyelid disease screening. Eyelid examination was performed by ophthalmology residents under the supervision of a specialist subspecializing in oculoplastics to identify any eyelid diseases (dermatochalasis, ptosis, entropion, ectropion, lash misdirection, presence of eyelid masses, and blepharitis). The prevalence of these eyelid diseases and their association with age and sex were determined.

Results: The 165 participants (122 females and 43 males) were all Chinese and were aged 24 to 92 (mean, 68) years; 23.6% were <60 years old, 62.4% were 60-80 years old, and 13.9% were >80 years old. The most common eyelid disease was dermatochalasis, with a prevalence of 61.8%, followed by blepharitis (32.1%), ptosis (7.3%), and eyelid mass (6.7%). Lash misdirection, entropion, and ectropion were uncommon, with a prevalence of 2.4%, 0.6%, and 0%, respectively. Dermatochalasis was more common with increasing age. Other eyelid diseases did not correlate with age.

Conclusion: Eyelid diseases are not uncommon among the general population in Hong Kong, particularly dermatochalasis, ptosis, and blepharitis. Eyelid diseases may negatively impact patients functionally and psychosocially. Clinicians and the general public should be aware of these eyelid diseases.

Key words: *Blepharitis; Ectropion; Entropion; Eyelid diseases; Eyelid neoplasms*

Introduction

Common eyelid diseases include dermatochalasis, ptosis, entropion, ectropion, lash misdirection, presence of eyelid masses, and blepharitis (**Figures 1** and **2**). Eyelid diseases may affect visual function and quality of life. Correction of eyelid diseases significantly improves vision-related quality of life.[1-4]

In Australia, the Blue Mountains Eye Study reported that 3.9% of the study population had ectropion.[5] In Brazil, the prevalence of involutional entropion and ectropion in the elderly population was 2.1% and 2.9%, respectively.[6] In Manchester, England, the prevalence of ptosis was as high as 11.5% in the aged population. Similar studies have not been conducted in Hong Kong.

Many eyelid diseases are involutional in nature. The prevalence of eyelid diseases increases in an ageing population. This may increase the public healthcare burden. This study aimed to determine the prevalence of eyelid diseases among the adult population in Hong Kong, and to determine the association of eyelid diseases with age and sex.

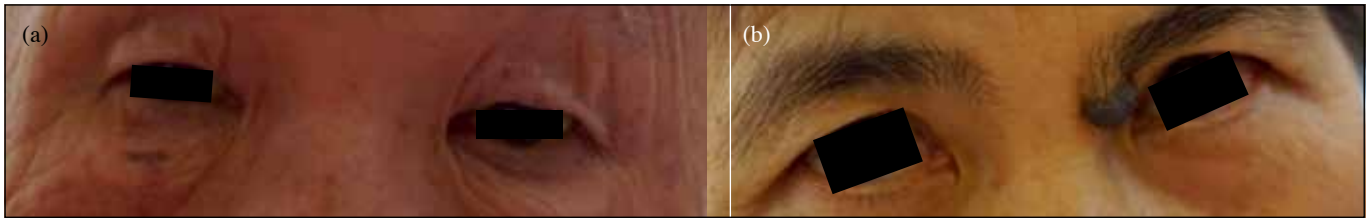

Figure 1. Clinical photographs showing (a) bilateral upper eyelid dermatochalasis and (b) a mass on the left upper eyelid.

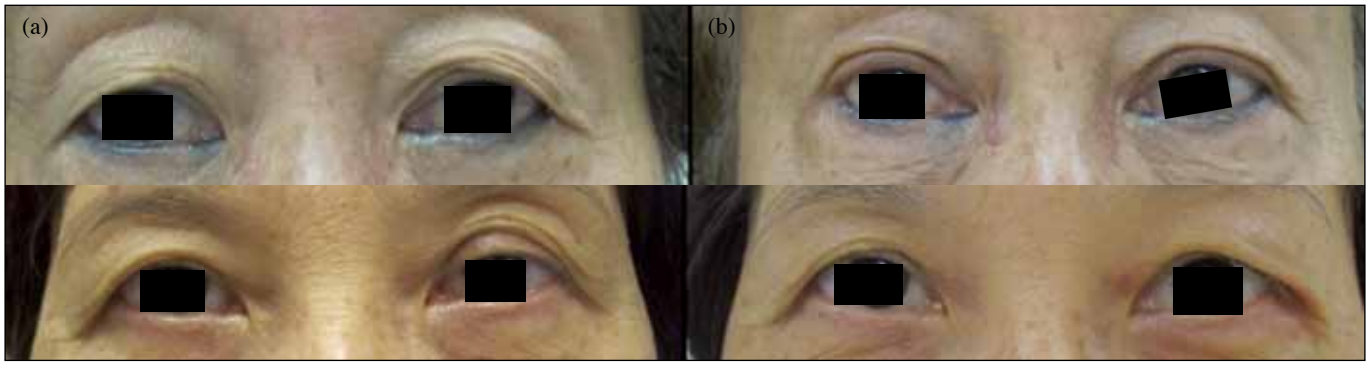

Figure 2. Clinical photographs before and after (a) blepharoplasty and (b) ptosis correction.

Methods

This cross-sectional study was approved by the Kowloon Central / Kowloon East Cluster Research Ethics Committee of the Hospital Authority (ref: KC/KE-18-0153-ER-4). On 22 October 2017, The College of Ophthalmologists of Hong Kong and The Hong Kong Ophthalmological Society jointly organized a Public Education Day (comprising health talks and eyelid disease screening) in a shopping mall in Tsuen Wan, New Territories, Hong Kong. A total of 165 participants were recruited for eyelid disease screening; about half were through the Hong Kong Association of Senior Citizens and the remaining by phone upon reading a recruitment advertisement in the Sky Post, a free newspaper. There was no age limit or sex restriction to participation.

Eyelid examination was performed by ophthalmology residents under the supervision of a specialist subspecializing in oculoplastics to identify any eyelid diseases (dermatochalasis, ptosis, entropion, ectropion, lash misdirection, presence of eyelid masses, and blepharitis). Dermatochalasis was defined as excessive eyelid skin on the upper eyelid hanging over the eyelashes.[7] Ptosis was defined as eyes with palpebral fissure height of <9 mm, or ≥2 mm difference between two eyelids.[8] Ectropion was defined as everted lower eyelid, whereas entropion as inverted eyelids. Lash misdirection can occur diffusely across the entire lid or in a small segmental distribution. Eyelid masses can occur anywhere in the upper or lower eyelids. Blepharitis was characterized by chronic inflammation of the eyelid, usually at the base of the eyelashes, with inflamed and reddened eyelids.

The prevalence of these eyelid diseases was determined. Association of these eyelid diseases with age and sex was determined using Chi-squared test. Statistical analysis was performed using the Statistical Package for the Social Sciences version 18.0 (SPSS, Chicago [IL], USA). A p value of <0.05 was considered statistically significant.

Results

The 165 participants (122 females and 43 males) were all Chinese and were aged 24 to 92 (mean, 68; standard deviation, 13) years; 23.6% were <60 years old, 62.4% were 60-80 years old, and 13.9% were >80 years old (**Table**).

The most common eyelid disease was dermatochalasis, with a prevalence of 61.8% (n=102). It occurred more commonly with increasing age, with prevalence of 25.6%, 71.8%, and 78.3% in the age-groups of <60 years, 60-80 years, and >80 years, respectively (p<0.001). The prevalence was comparable in males and females (58.1% vs. 63.1%, p=0.56).

Blepharitis was identified in 53 participants, giving a prevalence of 32.1%. The prevalence was not associated with age: 25.6%, 37.9%, and 17.4%, in the age-groups of <60 years, 60-80 years, and >80 years, respectively (p=0.10), and was comparable in males and females (34.9% vs. 31.1%, p=0.65).

Ptosis was identified in 12 participants, giving a prevalence of 7.3%. It occurred more commonly in males than females (18.6% vs. 3.3%, p=0.01). It occurred most commonly in

Table. Prevalence of eyelid diseases in the study population

Parameter	Prevalence (%)						
	Dermatochalasis	Ptosis	Entropion	Ectropion	Lash misdirection	Eyelid mass	Blepharitis
Overall (n=165)	61.8	7.3	0.6	0	2.4	6.7	32.1
Sex							
Male (n=43)	58.1	18.6	0	0	2.3	9.3	34.9
Female (n=122)	63.1	3.3	0.8	0	2.5	5.7	31.1
Age-group, y							
<60 (n=39)	25.6	10.3	0	0	5.1	7.7	25.6
60-80 (n=103)	71.8	5.8	1.0	0	1.0	4.9	37.9
>80 (n=23)	78.3	8.7	0	0	4.3	13.0	17.4

those aged <60 years (10.3%), followed by those aged >80 years (8.7%) and 60-80 years (5.8%) [p=0.64].

Eyelid masses were identified in 11 participants, giving a prevalence of 6.7%. They occurred most commonly in those aged >80 years (13.0%), followed by those aged <60 years (7.7%) and 60-80 years (4.9%) [p=0.34]. The prevalence in males and females was 9.3% and 5.7%, respectively (p=0.42).

Lash misdirection, entropion, and ectropion were uncommon, with a prevalence of 2.4%, 0.6%, and 0%, respectively.

Discussion

In our study, the prevalence of dermatochalasis was 61.8%, which is higher than the 17.8% reported in a Rotterdam study of 5578 people.[9] Dermatochalasis occurred more commonly with increasing age and is a feature of the aging process of the skin.[10] Male sex is a risk factor.[7] In our study, the prevalence of dermatochalasis was similar between males and females (58.1% vs 63.1%, p=0.56).

Dermatochalasis and ptosis have a negative impact on function and psychosociology.[11,12] Functionally, patients may experience superior visual field loss, impaired reading and other close-work ability owing to down-gaze ptosis, a chin-up backward head tilt owing to visual axis obscuration, symptoms of discomfort or eye strain owing to droopy lids, and in severe cases, interference with central visual owing to upper eyelid position.[11] Psychosocially, patients with blepharoptosis or dermatochalasis are viewed negatively.[12] Ptosis correction and blepharoplasty significantly improve vision-related quality of life,[1-4] with good functional and cosmetic outcome.

In our study, the prevalence of dermatochalasis and ptosis was 61.8% and 7.3%, respectively. The conditions may be undiagnosed in a large proportion of the population. Clinicians should proactively identify such patients and refer them to an ophthalmologist for further management. In our study, the prevalence of eyelid mass was 6.7%. Eyelid masses can be benign or malignant. In a review from Korea, approximately 36% of all eyelid tumors were malignant.[13] The incidence of malignant eyelid tumors has increased worldwide.[14-16] This trend was also reported in a local study, with the incidence increased from 0.6 per million people in 1997 to 2.3 per million people in 2009.[17] Biopsy should be performed to confirm the nature of the lesions. In our study, the prevalence of blepharitis was 32.1%. In a Korean study of the epidemiology of blepharitis in a large population-based sample, the prevalence was 8.1% among subjects aged ≥40 years.[18] Although blepharitis is often self-limiting, it can be severe or chronic and leads to lid malposition such as ectropion, keratitis, or corneal ulceration with consequent vision loss.[19] A Taiwan study reported that patients with blepharitis were at higher risk of developing anxiety and depression.[20] Blepharitis is associated with dysfunctional tear production or dry eyes and meibomian gland dysfunction. Ophthalmologist awareness of such conditions has increased since the publication of diagnosis and treatment guidelines for such disorders.[21] Clinical improvement can be achieved with simple measures such as cleaning the lid with diluted baby shampoo.[22] Other effective treatments include tear supplements and lubricants, various types of drugs, and nutritional supplements.[21] Given the high prevalence of blepharitis among the local population, more education should be provided to enable early identification of the disease to prevent possible complications and reduce the public healthcare burden.

Eyelid diseases can also affect other parts of the eye and body. For instance, untreated entropion can cause corneal abrasion, microbial keratitis, corneal vascularization, and subsequently vision loss.[23] Trachoma may cause lash misdirection and remains a leading infectious cause of blindness according to the World Health Organization.[24] Malignant eyelid masses, apart from being locally invasive, may result in lymph node metastasis, systemic metastasis, and even death.[25] These examples highlight the importance of eyelid health. Clinicians should refer patients with eyelid diseases to ophthalmologists for further assessment. Patients with complicated eyelid diseases should be managed by

oculoplastic subspecialists who are dedicated to eyelid, lacrimal, and orbital care.[26]

Our study has limitations. Recruitment of participants for screening was non-randomized, and hence there may have been selection bias. All participants were Chinese and therefore the results may not be directly comparable or applicable to other ethnic populations.

Conclusion

Eyelid diseases are not uncommon among the general population in Hong Kong, particularly dermatochalasis, ptosis, and blepharitis. Eyelid diseases may negatively impact patients functionally and psychosocially. Both clinicians and the general public should be aware of these eyelid diseases.

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Declaration

The authors have no conflicts of interest to disclose.

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CASE REPORT

Intraosseous hemangioma at the left lateral canthus: a case report

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Abstract

A 56-year-old Chinese woman presented with a 7-year history of a progressive, painless swelling at the left lateral canthus. Contrast-enhanced computed tomography (CT) of the orbit revealed a hypodense lesion of 1.4 cm at the anterior aspect of the left zygomatic arch. A radiological diagnosis of fibrous dysplasia was made. Debulking of the left lateral wall was performed. Intraoperatively, a diamond burr was used to smooth the surface and resulted in moderate bleeding, which was uncommon for fibrous dysplasia. Hemostasis was achieved with bone wax and direct compression. The pathological diagnosis was an intraosseous hemangioma.

Case Report

In October 2014, a 56-year-old Chinese woman presented with a 7-year history of a progressive, painless swelling at the left lateral canthus. Clinical examination revealed a non-tender mass with firm attachment at its base, of firm-to-hard consistency with no overlying skin change. Ophthalmic and neurological examination was unremarkable. Contrast-enhanced computed tomography (CT) of the orbit revealed a hypodense lesion of 1.4 cm at the anterior aspect of the left zygomatic arch, near the lateral wall of the left orbit (**Figure 1**). A radiological diagnosis of fibrous dysplasia was made.

In July 2017, debulking of the left lateral orbital wall was performed. Intraoperatively, an elevated lesion adhering to the lateral orbital rim was seen. A diamond burr was used to smooth its surface and resulted in moderate bleeding, which was uncommon for fibrous dysplasia. Hemostasis was achieved with bone wax and direct compression (**Figure 2**). The specimen was sent for pathological examination. Microscopy revealed medium-sized and dilated blood vessels lined with benign endothelium with poorly formed thin muscle coat in the stroma with reactive bones at the periphery. The pathological diagnosis was an intraosseous hemangioma at the left lateral orbital wall.

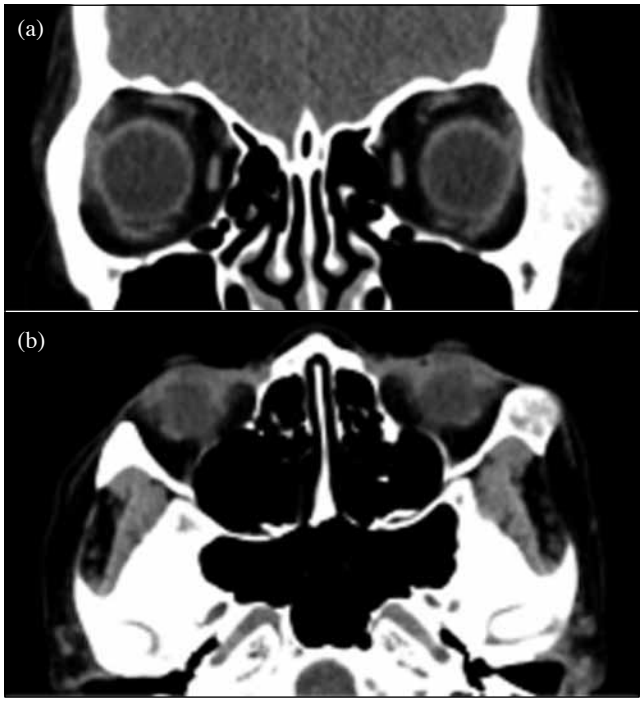

Figure 1. (a) Coronal and (b) axial contrast-enhanced computed tomographic scans of the orbit showing a well-circumscribed hypodense lesion at the anterior aspect of the left zygomatic arch with a sunburst and honeycomb appearance, near the lateral wall of the left orbit.

Figure 2. Hemostasis is achieved with bone wax and direct compression.

Discussion

Intraosseous hemangioma is rare and accounts for <1% of all bony tumors; its incidence is 2- to 3-fold higher in women than men.[1] Orbital intraosseous hemangiomas most commonly occur superiorly (35.3%) and inferolaterally (23.5%); only 5.9% occur laterally, as in our case.[2] Although the lesion is slow-growing and non-tender, posterior extension to the middle cranial fossa may cause a significant pressure effect with consequent vision loss, diplopia, or proptosis. On CT, the hypodense lytic configuration has a 'sunburst' or 'honeycomb' appearance. In our patient, retrospective review of the CT scans demonstrated the sunray pattern of trabeculation with well-circumscribed rarefraction (Figure 1). This made the diagnosis of intraosseous hemangioma probable.

Differential diagnoses of intraosseous hemangioma, in addition to fibrous dysplasia, include lytic bone lesions such as aneurysmal bone cyst, giant cell tumor, Langerhans cell histiocytosis, and osteogenic sarcoma.[3] Biopsy is essential to establish the diagnosis. The mainstay of treatment is *en bloc* excision with a small bone margin, with particular care to achieve hemostasis as the lesion is highly vascular. In our patient, hemostasis was achieved with bone wax and direct compression. Preoperative embolization is advocated if severe bleeding is expected.[4] In patients who are unfit for surgery or in lesions that are difficult to access, radiotherapy can be an alternative.[5] Recurrence after *en bloc* excision is uncommon.

Declaration

The authors have no conflicts of interest to disclose.

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NOVEL TECHNIQUE

Use of temporary safety sutures during intrascleral fixation of intraocular lens

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Sutureless intrascleral intraocular lens (IOL) implantation is an advanced scleral fixation technique to eliminate the suture-related complications of erosion and exposure after conventional methods.[1] Nonetheless, it is technically challenging. Crushing and pulling of the haptic of a three-piece IOL may cause deformation or breakage and hence tilting or dropping of the IOL. We introduce an additional step to enhance safety of the procedure.

Two additional 1.0-mm incisions are made at the peripheral cornea at 2 and 8 o'clock positions (**Figure a**). With the aid of a hook, a 10-0 nylon suture is inserted through each of the 1.0 mm paracenteses to form two suture loops in the anterior chamber (**Figure b**). The suture loop from the 8 o'clock incision is then externalized through the superior incision (**Figure c**). During insertion of the IOL injector, the cartridge is inserted through the externalized suture loop (**Figure d**). The leading haptic is then passed through the suture loop at the 2 o'clock incision during the injection process (***Figure e***). A pair of forceps is used to tighten the suture loop. After removal of the IOL injector, the trailing haptic is kept outside the eye to prevent the IOL from falling into the vitreous cavity. The trailing haptic is then entwined by the externalized suture loop originating from the 8 o'clock incision (**Figure f**). A pair of forceps is used to pull the suture loop back into the eye securing the haptic-optic junction of the trailing end (**Figure g**), followed by externalization of the leading and trailing haptics through the sclerotomy (**Figures h** to **j**). The two safety sutures enhance the stability of the IOL. After fixation of the externalized haptics, the two suture loops are removed.

The use of safety sutures during intrascleral IOL fixation is very helpful. In one case, the leading haptic was accidentally inserted posterior to the iris plane after the IOL was released from the injector. The IOL was prevented from dropping posteriorly by the safety suture at the 2 o'clock incision, which also aided manipulation of the leading haptic back to the anterior chamber. In another case, the leading haptic was bent and could not be inserted into the needle for externalization when performing the Yamane technique.[2] With the aid of the safety sutures, the IOL was stabilized in the anterior chamber while the bent leading haptic was rotated out of the main wound for trimming. The use of safety sutures is particularly helpful in cases of well-dilated pupil and aniridia.

In a series of 208 eyes undergoing sutureless scleral fixated IOL, intraoperative haptic breakage and deformation occurred in one and two eyes, respectively.[3] Only one of eight eyes undergoing sutureless scleral fixation had haptic breakage.[4] Manipulation of the trailing haptic can be difficult after the leading haptic is externalized. The leading haptic may rotate and slip back into the eye. Three cases of intraoperative haptic dislocation using the transconjunctival trochar-assisted technique have been reported.[5] The trailing haptic may be damaged when it becomes the only pivot point being held by the forceps to prevent dropping of the IOL posteriorly. A push-and-pull hook technique has been described to facilitate manipulation of the trailing haptic.[6] Keeping the leading haptic and holding needle inside the eye is suggested to facilitate externalization of the trailing haptic.[2]

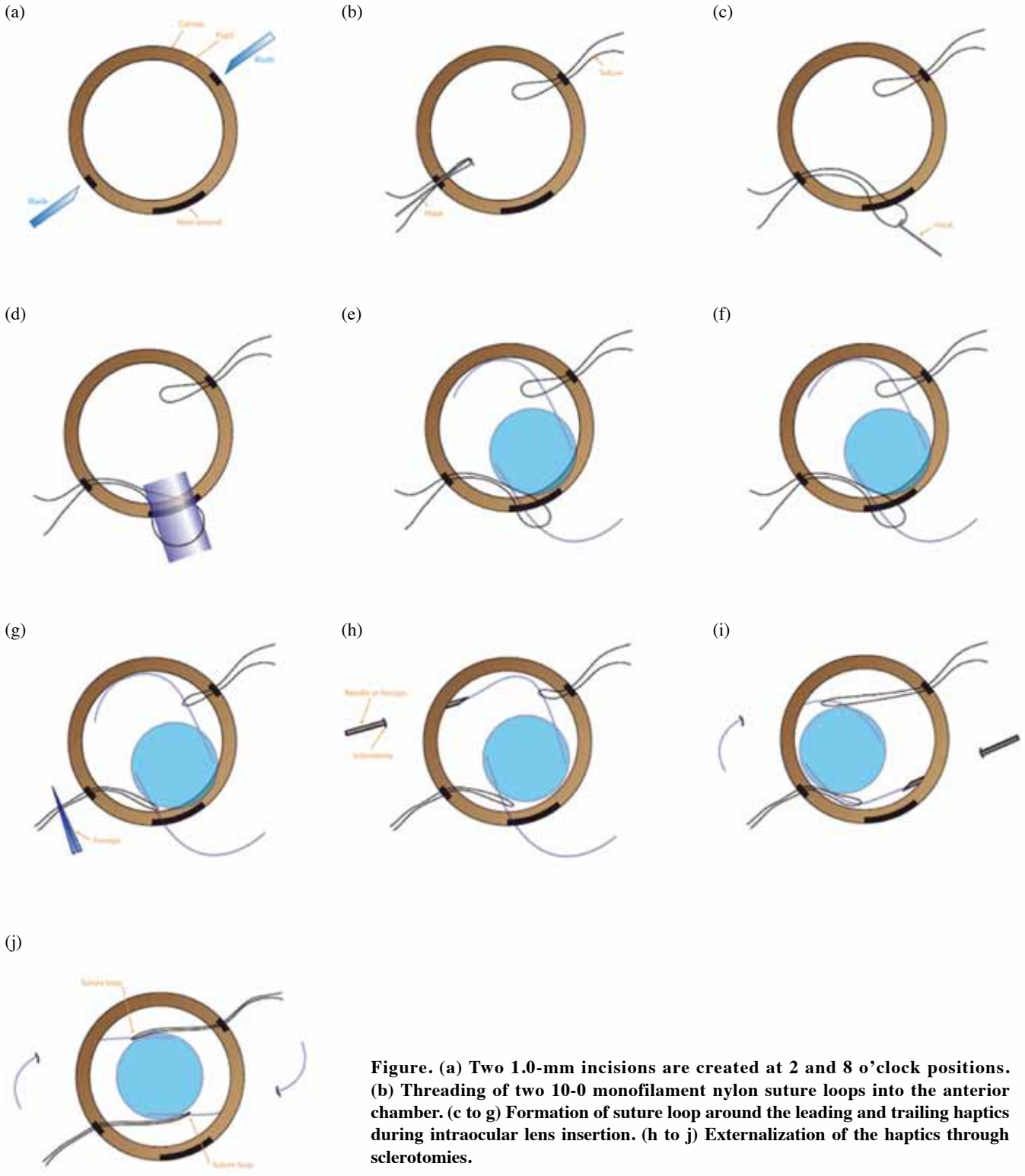

Figure. (a) Two 1.0-mm incisions are created at 2 and 8 o'clock positions. (b) Threading of two 10-0 monofilament nylon suture loops into the anterior chamber. (c to g) Formation of suture loop around the leading and trailing haptics during intraocular lens insertion. (h to j) Externalization of the haptics through sclerotomies.

Sutureless intrascleral fixated IOL implantation is increasingly popular among anterior segment surgeons. The IOL should stay within the sulcus and pars plana during the procedure after thorough anterior vitrectomy. Manipulation of the IOL can be stressful, especially when vitreoretinal support is not available. We added a maneuver to help stabilize the IOL. The technique is versatile and can be incorporated into various sutureless scleral fixation techniques and simple because it involves only the threading of two temporary sutures without tying knots. Although the use of safety sutures may be unnecessary for experienced surgeons, we recommend its use by trainees to enhance the safety of the procedure.

Declaration

The authors have no conflicts of interest to disclose.

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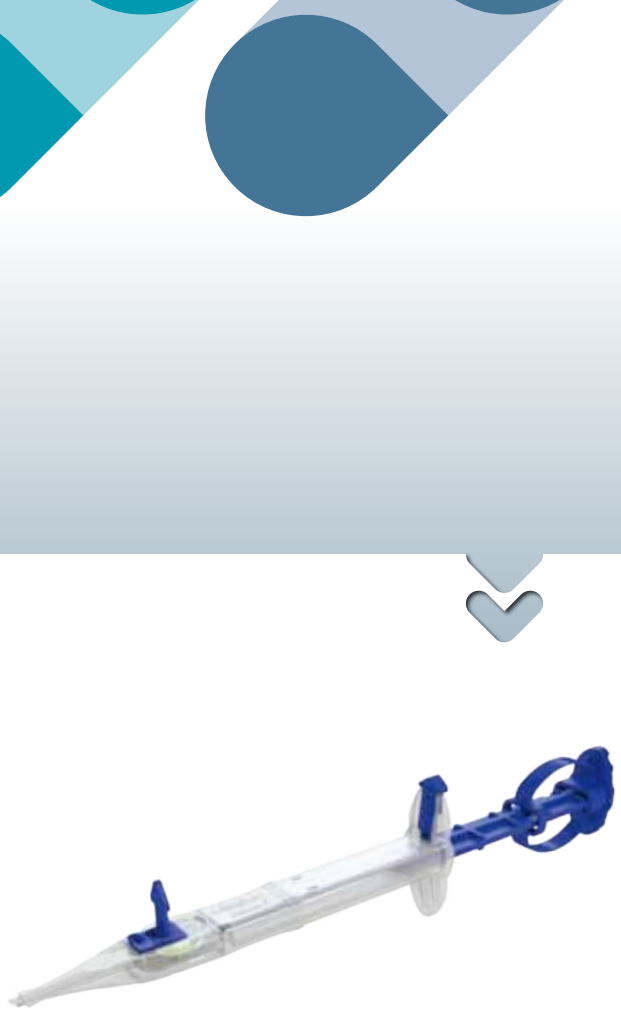

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