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Causes and Coping with Visual Impairment and Blindness

Edited by Shimon Rumelt

CAUSES AND COPING WITH VISUAL IMPAIRMENT AND BLINDNESS

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Notice

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Meet the editor

Prof. Shimon Rumelt received his medical degree and diploma in ophthalmology from Tel Aviv University, Israel. He completed his ophthalmology residency program at Western Galilee-Nahariya Medical Center in Nahariya, Israel; then an oculoplastics fellowship at Massachusetts Eye and Ear Infirmary, Boston, MA; and a vitreoretinal fellowship at Boston University, MA. He earned a master's degree in Public Administration (Health Systems) from Clark University, Worcester, MA. He is a senior ophthalmologist at the Western Galilee-Nahariya Medical Center and is engaged with various fields in ophthalmology. His interests include clinical activities, surgery, research, and teaching medical students, residents, and fellows. Prof. Rumelt has edited five books and is an author and a coauthor of approximately 100 scientific articles and book chapters. He is a member of the editorial board of *Evidence-Based Ophthalmology* and a reviewer for multiple professional journals. He is an associate clinical professor at the Faculty of Medicine, Bar-Ilan University, Zefat, Israel.

Contents

Preface

Blindness and cancer are the two most feared entities worldwide. The World Health Organization (WHO) estimated that 285 million people (4%) out of the 7.2 billion world population had either low vision (246 million) or blindness (49 million) in 2014. Ninety percent of them live in low-economic settings and eighty-two percent are over 50 years of age. Eighty percent of visual impairment can be prevented or cured. The best examples are correction of refractive errors and cataract surgery. Although 230,000 ophthalmologists are spread throughout the world, their distribution is uneven. Even if it were even, lack of functional ophthalmic instruments and medications in many parts of the world prevents proper treatment. Thus, the essential purpose of ophthalmologists and researchers is to prevent blindness and, if it is reversible, to cure it. Indeed, patients should leave the ophthalmology department or clinic better than when they came in. The degree of improvement may be variable and we as ophthalmologists should aim for the best. When patients leave the department or clinic worse than when they came in, we have failed. Although most of the common eye diseases that impair vision can be treated fairly simply, for some diseases, the cure may be limited and early diagnosis and treatment are imperative (e.g., retinal disorders and glaucoma). In case of inevitable impaired vision or blindness, successful coping is required from the patients, their families, surroundings, society, and the state because blindness is a major burden for all.

This book is aimed at addressing different causes of visual impairment and blindness, their epidemiology, manifestations, risk factors, prevention of progression, and treatment. It is aimed at encouraging physicians and researchers to increase efforts to prevent irreversible and treat reversible blindness for the betterment of the world. Therefore, it is essential to be fully aware and knowledgeable of the manifestations of the diseases causing blindness and this book covers some of their different aspects. Each chapter was written by experts from around the globe. Thus, it reflects the importance of the subject.

The book is divided into three sections. The first section, Causes of Blindness, includes chapters on blindness due to mustard gas neuro-ophthalmological diseases and glaucoma. The second section addresses the evaluation and manifestation of visual impairment (glare in ocular disorders). The third section, Coping with Visual Impairment and Blindness, contains chapters on coping with impairment, psychological adaptation, assistive systems, and designing hands-on robotic courses.

My sincere and deep gratitude goes to the authors for their time and effort. Deep appreciation goes to Ms. Romina Rovan, the Publishing Process Manager, for her endless devotion to publishing this book, and to the publisher IntechOpen for this excellent project. This book is a balanced result of efforts to publish in a timely manner and to cover the topic as much as possible. Hopefully, additional books will cover more aspects of this important issue.

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Section 1

Causes of Blindness

Chapter 1

Corneal Blindness Caused by Mustard Gas

Sepehr Feizi

Additional information is available at the end of the chapter

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Abstract

Mustard gas is a lipophilic, highly cytotoxic agent that rapidly penetrates tissue, and the eye is one of the organs mostly affected. Mustard gas-related ocular injuries can be divided into immediate, chronic, and delayed-onset phases. Late complications, developing after 1–40 years, can cause progressive and permanent reduction in visual acuity and even blindness. A wide range of late ocular involvements have been reported, which include chronic blepharitis, limbal ischemia and stem cell deficiency, and corneal scarring and neovascularization. The majority of corneal involvements are limited to the anterior stroma, leaving the posterior stroma and endothelium relatively intact. Therefore, lamellar keratoplasty is appropriate for the management of corneal involvements in the majority of victims. This procedure can be performed alone or in combination with limbal stem cell transplantation.

Keywords: corneal blindness, mustard gas, corneal involvement, keratitis

1. Introduction

1.1. History

The exact date of the first sulfur mustard synthesis is somewhat unclear, but the first report by Despretz may have been in 1822. An 1860 report by Neimann described a delayed-effect vesicant oil as a reaction product of ethylene on a mixture of sulfur chlorides. In 1886, a process to produce significant quantities of pure sulfur mustard was described by Meyer using sodium sulfide, ethylene chlorohydrin, and hydrochloric gas [1, 2].

Mustard gas was used for the first time by German forces against Allied troops in July 12, 1917 that caused more than 2100 casualties. The Allies began using mustard gas against German troops in 1918. During 1935–1936, the Italian army dropped mustard-gas bombs in Ethiopia to destroy Emperor Haile Selassie's army. During 1963–1967, Egypt used mustard gas and a

nerve agent in Yemen to support a coup against the Yemeni monarchy. During the Iran-Iraq war (1980–1988), Iraq used chemical weapons, including tabun and mustard gas, against Iran and Iraq's Kurdish minority. Iraq's use of chemical weapons was confirmed by the United Nations experts [3].

1.2. Molecular formula of mustard gas and its biochemical mechanism of tissue injury

Sulfur mustard ($C_4H_8Cl_2S$) is one of a class of chemical warfare agents which are known as vesicants because they cause vesicles, or blisters, on exposed skin. Pure sulfur mustard is odorless, colorless, and viscous liquid at room temperature. It is usually yellow-brown in color and has an odor resembling garlic, horseradish, or mustard plants when used as warfare agents, which is how it got its name. However, this compound has absolutely no relation whatsoever to culinary mustard [4].

Mustard gas is a lipophilic, highly cytotoxic agent that rapidly penetrates tissue [5]. Exposed skin surfaces, eyes, the linings of both respiratory and gastrointestinal tracts, and renal systems as well as the bone marrow are all at risk. The risks increase dramatically under hot, humid conditions, and it can be lethal at sufficiently high doses [5, 6]. It has been demonstrated that 80% of sulfur mustard applied to the skin evaporates, 10% remains in the skin, and 10% gets absorbed systemically [7]. Susceptibility of the eyes to the toxic effects of mustard gas is due to moistness of the ocular surface, allowing activation of the agent. Additionally, corneal epithelial cells have a high turnover and metabolic rate that increase their vulnerability to the lipophilic sulfur mustard trapped into the oily tear layer [8].

Sulfur mustard is a cellular poison that triggers apoptosis as a cytotoxic mechanism. The acute toxic effects of mustard vesicants are usually attributed to the consequences of alkylation reactions with organic compounds including nucleoproteins such as DNA [9]. The ladder pattern of DNA fragmentation after cell exposure to mustard gas indicates internucleosomal cleavage of DNA. Alkylation reactions can result in genotoxic effects as well as physiological and metabolic disturbances that induce apoptosis [10]. In addition, mustard gas is a mutagen and is a known carcinogen that is associated with an increased risk of developing lung and other respiratory tract cancers [11].

2. Mustard gas-related ocular injuries

Mustard gas-related ocular injuries can be divided into immediate, chronic, and delayed-onset phases [12]. Acute manifestations of varying degrees, including eyelid erythema and edema, chemosis, subconjunctival hemorrhage, epithelial edema, punctate erosions, and corneal epithelial defects, develop in 75–90% of exposed individuals and can follow three different courses: complete resolution, persistent smoldering inflammation (chronic form), or reappearance of lesions after a latent period of quiescence (delayed form) [13, 14].

Late complications, developing after 1–40 years, can cause progressive and permanent reduction in visual acuity and even blindness, and they occur in approximately 0.5% of those initially

severely wounded [6, 13]. A wide range of late ocular involvements have been reported, which include chronic blepharitis, dry eye, conjunctival vessel tortuosity, limbal ischemia and stem cell deficiency, corneal scarring and neovascularization, corneal thinning and perforation, epithelial irregularity, recurrent or persistent epithelial defects, and secondary degenerative changes including lipid/amyloid deposits (**Figure 1**) [5, 6, 12–17].

Dry eye is a late ocular complication of exposure to mustard gas, the symptoms of which are often severe and persistent and can influence many aspects of intoxicated victims' lives [5, 6, 18, 19]. Although the exact pathophysiologic cause of dry eye syndrome after exposure to mustard gas is not known yet, most studies in this regard have revealed evidence for increased apoptosis in the conjunctival epithelium [20]. This apoptosis also occurs in goblet cells resulting in a significant decrease in goblet cell density thus reducing mucin production and tear film stability [20]. Additionally, dysfunction of lacrimal glands may occur secondary to lymphocytic infiltration of the glands [20].

Mustard gas-related corneal involvements are completely different from those observed in other causes of corneal opacities that develop after trauma, infection, and acid or alkaline burns [18]. For example, corneal thinning and fragility is a striking feature in mustard gas-induced ocular injuries [18]. Such differences can be explained by the presence of other concomitant abnormalities such as limbal ischemia and vascular abnormalities [18]. Limbal ischemia causes scleral and corneal thinning, and the presence of leaking limbal vessels results in the accumulation of abnormal materials such as lipid and amyloid in the adjacent cornea [12]. Alterations of corneal stroma secondary to acute and chronic inflammation, stromal scar and fibrosis, and deposits make stromal layers too rigid to be separated by air. Therefore, deep anterior lamellar keratoplasty using the big-bubble technique is hard to perform in mustard gas-induced keratitis [12].

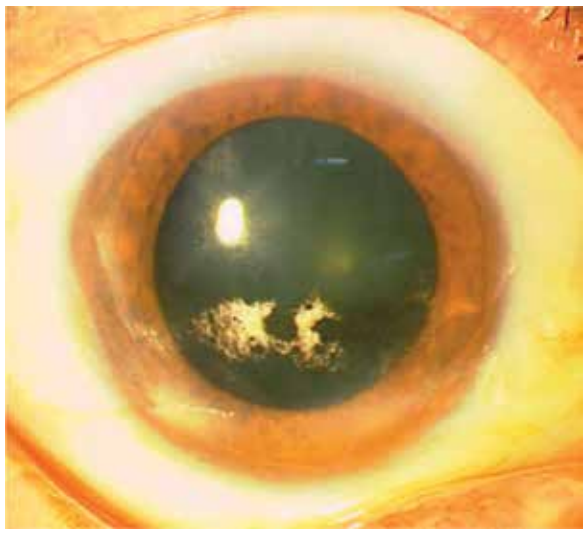

Figure 1. Abnormalities of the cornea, including surface irregularity, thinning, and intrastromal lipid and amyloid deposits, are evident in an eye suffering from mustard gas keratitis.

Although limbal stem cell deficiency has been reported in mustard gas-related ocular involvements, its clinical manifestations are completely different from those observed in other causes of stem cell deficiency such as acid or alkaline burns, thermal burns, Stevens–Johnson syndrome, ocular cicatricial pemphigoid, and multiple surgeries [18]. For example, conjunctivalization of the corneal surface, which is a striking feature in the latter conditions, is hardly observed in mustard gas-induced keratitis. Additionally, there is no correlation between the severity of corneal involvements and limbal stem cell deficiency in these eyes [12]. Limbal abnormalities observed in mustard gas-induced ocular involvements are contributed by the combined effects of limbal stem cell deficiency, limbal ischemia, and abnormally leaking vessels. However, one mechanism can be more prominent than the others in certain cases [12].

3. Management of mustard gas-induced ocular involvements

3.1. Management of acute phase

The management of the acute phase is relatively straightforward, chiefly consisting of symptomatic therapy to address the patient's discomfort and ocular inflammation. This approach includes copious irrigation with potable water at the time of exposure, topical antibiotics, preservative-free lubricants, and anti-inflammatory agents [21]. Artificial tears and lubricating ointments should be administered every 6 hours. Topical antibiotics (e.g., chloramphenicol or ciprofloxacin eye drops) should be prescribed every 6 hours for 7–10 days to prevent bacterial infection [21]. Topical steroids and non-steroidal anti-inflammatory drugs are found to be beneficial in ameliorating the initial inflammatory response and in postponing the development of corneal neovascularization. Corticosteroid eye drops should be administered every 8–12 hours for a week and then gradually tapered over 2–3 weeks [21]. The prolonged use of topical corticosteroids (more than 3 weeks) should be avoided. Amniotic membrane transplantation can be considered for the management of acute phase because it suppresses inflammation and scarring and promotes healing [18]. Symblepharon formation is not the feature of mustard gas-induced ocular involvements. Therefore, the victims do not require symblepharolysis [12].

3.2. Management of chronic phase

To date, there is no definitive therapy for chronic and delayed-onset mustard gas-related keratitis. Therapy for delayed phase is tailored on the basis of the severity and type of involvements and can vary from symptomatic therapy to surgical interventions for ocular surface problems including dry eye and corneal epithelial instability, corneal opacity, and limbal stem cell deficiency [12].

3.2.1. Medical managements

Different medications have been used for the management of sulfur mustard-induced ocular injuries. They include preservative-free artificial tears, topical steroids and antibiotic, N-acetylcysteine, topical cyclosporine A, resolvin E1, topical form of essential fatty acids, thymosin β4, topical form of curcumin, newly formulated artificial tears, diquafosol, rebamipide, tretinoin, and oral uridine.

Preservative-free artificial tears and lubricants are one of the most prescribed drugs in the management of ocular symptoms in mustard gas-related corneal involvements. New formulas have been proposed for artificial tears, and can be used in the management of dry eye disease with any etiology. Recombinant human lubricin (proteoglycan 4), a natural substance [22], and hyaluronic acid with trehalose [23] are one of these new formulas, and have been found to be safe with a better patient satisfaction. Natural components of tear film such as anionic glycosaminoglycan polysaccharide in combination with polymers, hyaluronic acid, and carmellose sodium are quite effective in corneal epithelial staining [24]. Another new formula contains carmellose sodium, osmoprotectants, and hyaluronic acid and has been demonstrated to improve ocular symptoms in dry eye disease [25].

Curcumin is an anti-inflammatory agent with anti-cancer and anti-apoptotic properties [26, 27]. Dietary curcumin is found to decrease lens opacification in a rat model of naphthalene-induced cataract [28]. Curcumin is effective in the management of different respiratory and cutaneous symptoms in sulfur mustard-exposed casualties [29, 30]. This natural hydrophobic polyphenol is proposed as an alternative treatment for dry eye disease [31]. Maria et al. [32] have developed a formulated eye drop for curcumin with more aqueous solubility properties. This sustained-release drop may be appropriate for the management of different inflammatory ocular surface disorders encountered in sulfur mustard-exposed patients [32]. However, further animal studies and clinical trials are required to approve the efficacy of this formulation.

Resolvin E1 (RvE1), a derivative of eicosapentaenoic acid, is an endogenous lipid mediator and can inhibit pro-inflammatory responses [33]. This drug has been used for the management of periodontitis, inflammatory bowel disease, and prevention of vascular inflammation [33, 34]. It has been shown that the topical administration of RvE1 significantly down regulates cyclooxygenase-2 (COX-2) expression and increases tear production, resulting in an increase in the density of superficial epithelial cells in a dry-eyed mouse model [35]. These features make RvE1 a potential therapeutic option in delayed ocular lesions induced by sulfur mustard. Similarly, thymosin β4 eye drops have been found to be effective in the treatment of dry eye disease and corneal vascularization and thus may have a role in the management of delayed ocular lesion in sulfur mustard-exposed victims [36]. There is no report of ocular toxicity associated with the topical form of thymosin β4 [36].

Diquafosol is a P2Y2 purinergic receptor agonist that stimulates the receptors in ocular tissues and thus increases mucin (conjunctival goblet cells stimulation) and the aqueous portion of tear film (conjunctival epithelial cells stimulation) [37]. Three percent diquafosol ophthalmic solution is effective for the treatment of dry eye disease through tear film stabilization and repair of corneal epithelial damages [38, 39]. Another P2Y2 receptor agonist is uridine. Oral uridine is reported to be beneficial for increasing mucin secretion and tear production [40].

Rebamipide is a mucosal protective agent with anti-inflammatory, immunosuppressive, and anti-apoptotic activities [41]. Corneal and conjunctival mucin can be effectively and safely increased by 2% rebamipide ophthalmic suspension [42]. The efficacy of diquafosol, rebamipide, and oral uridine in sulfur mustard-exposed patients should be investigated in clinical trials.

Tretinoin (0.01% all-trans-retinoic acid) is effective in the treatment of dry eye disease [43]. Tretinoin improves tear film break-up time and Schirmer tear test results [43]. However, tretinoin cannot

improve ocular symptoms such as foreign body sensation and photophobia [43]. Therefore, it can be considered a secondary option for the treatment of mustard gas-induced dry eye. Tretinoin in combination with topical interferon α-2b is an option in the management of partial limbal stem cell deficiency [44].

Cyclosporine A is an approved immunomodulatory ophthalmic product with different concentrations (0.05, 0.1, 1, and 2%) and has been used to increase tear production in dry eye patients [45]. Cyclosporine blocks the IL-2 signaling pathway and then inhibits T cell-mediated immune response [46]. Recently, Jadidi et al. [20] demonstrated that the treatment with topical cyclosporine A 0.05% in patients with severe dry eye due to mustard gas injury increased goblet cell density in the bulbar conjunctiva and improved symptoms of the disease.

Omega-3 plays important roles in human biology through decreasing cytokines and inhibiting oxidative stress [47]. It is an anti-inflammatory agent that prevents apoptosis in ocular tissues and can reduce tear osmolality and increase tear production in patients with dry eye disease [48–50]. However, there is no evidence of its significant effectiveness in meibum lipid composition [50]. A topical form of omega-3 fatty acid in combination with hyaluronic acid could successfully treat dry eye in a mouse model [47].

N-acetylcysteine, a mucolytic agent, has been used in ophthalmology to prevent corneal melting and perforation in different corneal diseases, including alkali-burned corneal ulcers [51] and filamentous keratitis [52]. N-acetylcysteine is a derivative of cysteine, which inhibits collagenase irreversibly by reducing disulphide bonds and by chelating calcium or zinc. It also inhibits matrix metalloproteinase-9 (MMP-9), potentially by similar mechanisms. N-acetylcysteine may be useful clinically to treat corneal destruction in mustard gas-induced keratitis in which MMP-9 activity and inflammatory cytokines are upregulated [53]. Although the exact mechanism of inhibition of MMP-9 secretion by N-acetylcysteine is currently unknown, the inhibitory properties of N-acetylcysteine on inflammation has been shown to act through nuclear factor-kB, which has a pivotal role in inducing the expression of multiple genes in immune and inflammatory responses [54, 55]. Topical applications of 5 and 20% N-acetylcysteine have been shown to be effective in the treatment of ocular surface disorders without toxic effects. However, prolonged treatment of N-acetylcysteine has adverse effects on the cornea.

3.2.2. Surgical managements

Dry eye can be addressed surgically using punctal plug, punctal occlusion, and temporary or permanent tarsorrhaphy [12]. Amniotic membrane transplantation is an effective approach for the management of persistent epithelial defects when limbal stem cell deficiency is partial because it suppresses inflammation and scarring and promotes healing [18]. Amniotic membrane transplantation is beneficial with keratectomy in the case of lipid deposition, and it has been used as a graft in limbal stem cell transplantation [18]. However, transplanted amniotic membrane can integrate into the corneal stroma, resulting in a reduction in visual acuity (**Figure 2**).

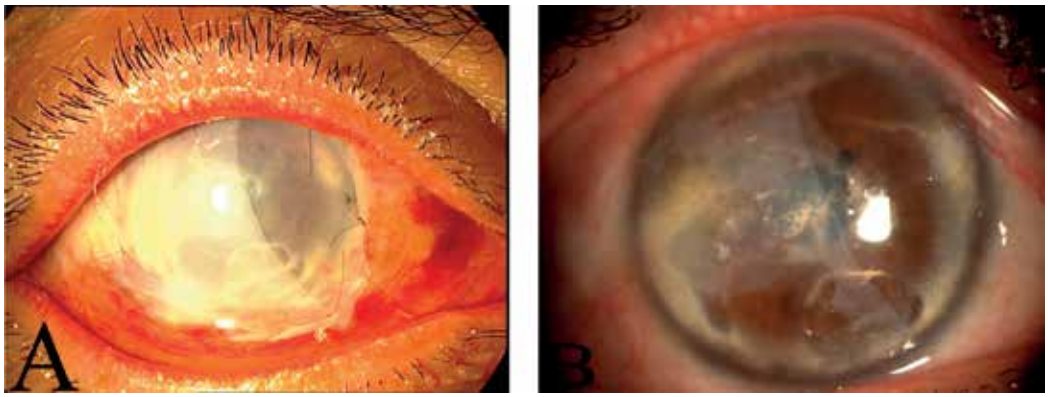

Figure 2. (A) Amniotic membrane transplantation was performed for the management of persistent epithelial defects in victim of mustard gas. (B) Please, note transplanted amniotic membrane has integrated into the corneal stroma, resulting in a reduction in visual acuity.

When significant limbal ischemia and/or stem cell deficiency develop, stem cell transplantation is required to provide a viable source of corneal epithelial cells as well as address conjunctival ischemia and scleral and peripheral corneal thinning [56, 57]. Limbal stem cells can be harvested from first-degree relatives, including parents or siblings (living-related conjunctival-limbal allograft) [56]. However, we noticed that living-related conjunctival-limbal allograft cannot provide adequate corneal and scleral lamellae, and cadaveric eyes should also be available [56]. Therefore, the technique of limbal stem cell transplantation was changed to keratolimbal allograft, which is harvested from cadavers and can provide more stem cells [57]. Another advantage worth mentioning is that keratolimbal allograft makes it possible to harvest corneal and limbal blocks from the same donor, if both transplantations are to be performed simultaneously [57]. This approach can reduce the antigenic load to the recipient's immune system [57].

3.2.2.1. Corneal transplantation for the management of mustard gas-induced keratitis

Traditionally, penetrating keratoplasty has been commonly performed as an ultimate treatment of different corneal pathologies, and numerous studies have reported good visual results after surgery [58]. We performed penetrating keratoplasty in 27 eyes of 27 victims of mustard gas and followed the patients for 15–96 months [58]. We reported a graft survival rate of 77.3%, indicating relatively acceptable outcomes, especially when corneal opacity is centrally located, and there is no severe limbal involvement (**Figure 3**) [58]. However, in cases demonstrating severe dry eye, limbal ischemia, or peripheral corneal involvements, a high rate of graft failure due to rejection reactions or recurrence of opacity was noted (**Figure 4**) [58]. Additionally, most corneal involvements are limited to the anterior stroma, leaving posterior stroma and endothelium relatively intact [59–61]. Therefore, the technique of corneal transplantation has evolved from penetrating keratoplasty to lamellar keratoplasty.

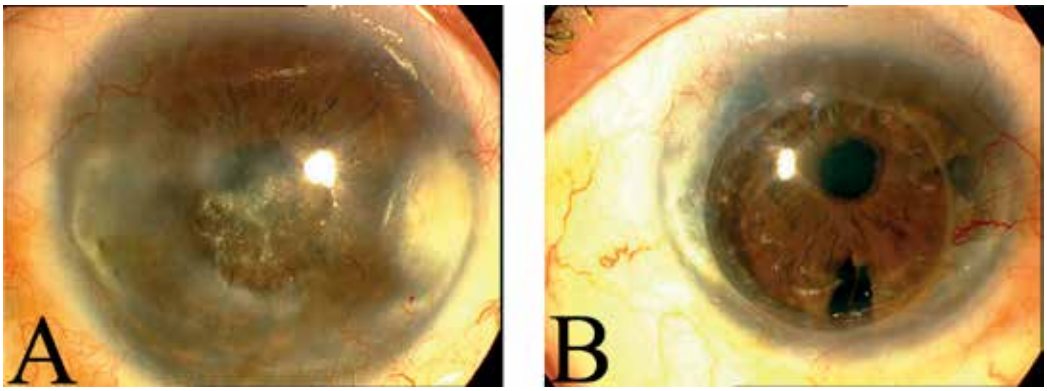

Figure 3. (A) Signs of mustard gas-induced keratitis, including surface irregularity, thinning, and intrastromal lipid and amyloid deposits, are evident. (B) Penetrating keratoplasty was performed in the same eye and yielded a clear graft 16 months postoperatively in an eye with mustard gas keratitis.

3.2.2.1.1. Indications for conventional lamellar keratoplasty

Indications for optical or tectonic lamellar keratoplasty in mustard gas-induced corneal involvements are corneal haziness leading to decreased visual acuity, photophobia, discomfort caused by corneal surface irregularity, abnormal deposits or severe corneal thinning threatening globe integrity, or a combination thereof [12]. A full-thickness graft is still inevitable in certain conditions such as significant interface opacity, deep stromal scar, and corneal perforation [12].

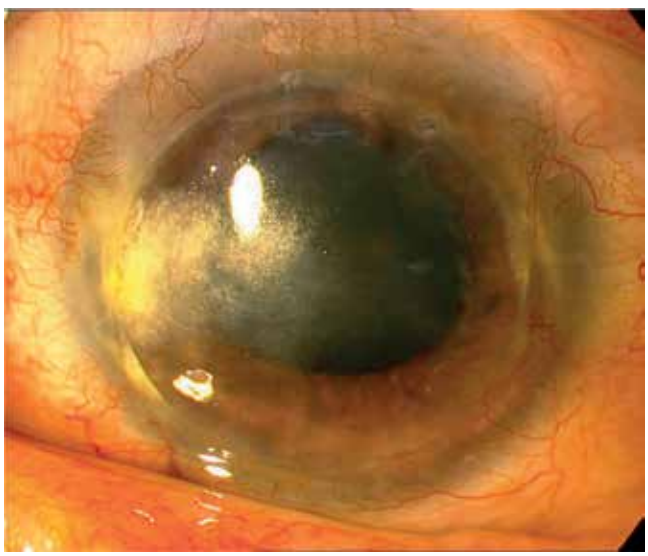

Figure 4. A penetrating keratoplasty graft opacity results from limbal stem cell deficiency and ocular surface abnormalities in a victim of chemical warfare.

3.2.2.1.2. Surgical technique

Manual lamellar dissection technique is used to perform conventional lamellar keratoplasty. The size of recipient trephine is selected on the basis of the vertical corneal diameter and the extent of corneal involvements. Based on the corneal thickness and the depth of opacities, at least 70% of the corneal thickness is trephined and manual lamellar dissection is performed using a crescent blade. During lamellar dissection, it is attempted to remove all deposits and opacities, mainly confined to the anterior- and mid-stroma, and to create a smooth, clear, and single-plane recipient bed. A partial-thickness donor corneal graft, prepared from a fresh whole globe with an intact epithelium, oversized by 0.5 mm, and matching the depth of the recipient bed is sutured using combined 8-bite interrupted sutures accompanied by 16-bite single running 10-0 nylon sutures (**Figure 5**). The suture tension should be moderate because very tight sutures prevent the appropriate spreading of tear film over the cornea, retarding reepithelialization of the graft in such compromised eyes.

We performed conventional lamellar keratoplasty in 51 eyes with mustard gas-induced corneal involvements and followed up the patients for 19–107 months. Best-corrected visual acuity was 0.35 ± 0.16 (0.0–0.48) logMAR at the final follow-up and graft survival rate was 91.7% in this series [58]. Postoperative complications included epithelial rejection (two eyes), persistent epithelial defects (four eyes), graft opacity (three eyes), and significant interface haziness (three eyes) [58].

3.2.2.1.3. Concomitant procedures

The unique features of mustard gas-induced ocular injuries are limbal and corneal involvements [12]. Based on the severity of dry eye, corneal epithelial instability, and

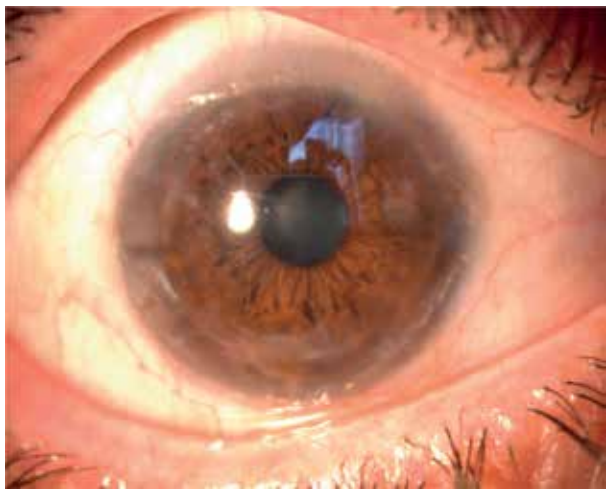

Figure 5. Lamellar keratoplasty was performed for mustard gas-induced keratitis. Despite the presence of Descemet's membrane wrinkling, the patient has acceptable visual acuity.

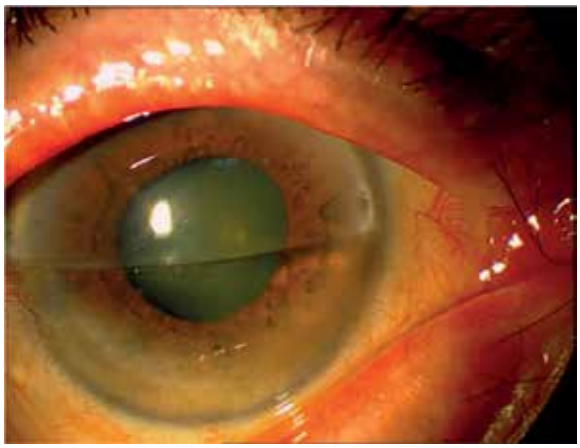

Figure 6. Simultaneous en bloc keratolimbal allograft and lamellar keratoplasty were performed to address coexisting mustard gas-induced limbal stem cell deficiency and corneal involvements in the same patient.

limbal stem cell deficiency, other interventions including punctal occlusion, temporary or permanent tarsorrhaphy, and limbal stem cell transplantations may be required to address these late complications at the time of corneal transplantation. A significant number of participants require both limbal stem cell and corneal transplantations. There is a trend to carry out both lamellar keratoplasty and limbal stem cell transplantation at a single session (**Figure 6**) to decrease the number of surgeries and anesthesia administrations [57]. In addition, only one donor can be used to provide both stem cells and cornea during a simultaneous operation, which can decrease the antigenic load presented to the recipient's immune system [57]. Furthermore, the total duration of oral corticosteroid and immunosuppressive treatment is shorter in the simultaneous approach than in the sequential approach, which require oral corticosteroid and immunosuppression after each surgical intervention [57].

3.2.2.1.4. Postoperative medical regimens

Postoperatively, the patients are medicated with 0.5% chloramphenicol eye drops every 6 hours, 0.1% betamethasone eye drops every 6 hours, preservative-free artificial tears every 2 hours, lubricating ointments every 8 hours, and systemic prednisolone 1 mg/kg daily [12]. Topical antibiotics are discontinued after complete epithelial healing, whereas oral and topical corticosteroids are tapered off over 2–4 weeks and 2–3 months, respectively, based on the severity of ocular inflammation [12].

For patients who undergo limbal stem cell transplantation concomitantly, systemic tacrolimus 1 mg twice a day is started at the time of surgery and continued for 1.5 years. Additionally, 1 g oral mycophenolate mofetil is prescribed twice daily for at least 6 months. It is then tapered gradually and discontinued after 1 year [12]. Cell blood counts, blood pressure, and renal and liver function test results should be monitored at appropriate intervals in collaboration with a kidney transplantation expert to monitor for possible complications of immunosuppressive therapy.

3.2.3. Complications of lamellar keratoplasty

3.2.3.1. Intraoperative complications

3.2.3.1.1. Descemet's membrane perforation

Perforation in Descemet's membrane during lamellar keratoplasty which is reported to be as high as 26.3% [62] can occur during any step of surgery including trephination, stromal excision, and donor suturing. The moment of perforation is crucial for the completion and the success of lamellar keratoplasty. Early perforations make stromal dissection more difficult and result in a greater residual stroma and hence slower visual recovery because of interface opacification [63]. Additionally, the size of perforation determines the severity of endothelial damage; a large hole within Descemet's membrane leads to a flat anterior chamber necessitating multiple air injections which is associated with more severe endothelial damage [63].

Management of Descemet's membrane perforation depends upon the location and size of the hole and the step of surgery at which this complication takes place. Perforations that occur during trephination can be managed by tight sutures at the site of perforation before lamellar dissection. Finally, sutures which fix donor tissue to the recipient bed in the site of perforation should be full thickness including recipient Descemet's membrane. If perforation occurs during stromal dissection, the site of perforation should be dissected last at a different plane to leave some stroma over this area to seal the perforation. At the end of operation, the anterior chamber is partially filled with an air bubble. If anterior chamber is completely filled with air, pupillary block may develop postoperatively. For perforation occurring during suturing, injection of air into the anterior chamber at the conclusion of surgery usually suffices.

3.2.3.2. Postoperative complications

3.2.3.2.1. Persistent epithelial defects

Because of ocular surface abnormalities including severe blepharitis and dry eye as well as limbal stem cell deficiency, victims of mustard gas are susceptible to develop persistent epithelial defects after keratoplasty [12]. Persistent epithelial defects can lead to postoperative complications such as subepithelial scarring or infectious keratitis. Therefore, it should appropriately be managed. Intraoperative measures include the use of good donor quality grafts with intact epithelium and the avoidance of damage to the corneal epithelium during the preparation of the donor tissue. Additionally, appropriate suture tension and surgical wound apposition, punctal occlusion, and tarsorrhaphy encourage the epithelialization of donor grafts [21]. Postoperatively, control of inflammation with topical steroids, deliberate use of preservative-free artificial tears and lubricating ointments as well as treatment of blepharitis are advisable [21]. Sometimes, it is necessary to prescribe autologous serum 20% and/or fit a bandage contact lens in intractable cases [21].

3.2.3.2.2. Suture-related complications

Suture-related complications such as sterile reactions, early suture loosening, cheese-wiring, and suture-related scarring and vascularization can develop after lamellar keratoplasty when

suture are in place. These complications can be reduced or even prevented by appropriate suture depth, length, and tension. Additionally, the administration of topical corticosteroid for an adequate period after surgery can significantly reduce suture-related complications. However, the long-term use of corticosteroid eye drops may increase the risk of cataract, glaucoma, graft ulcer, and endophthalmitis from micro-defects or microperforation near the suture.

3.2.3.2.3. Refractive error and astigmatism

Similar to penetrating keratoplasty, postoperative myopic and astigmatic refractive errors remain the main reason for patient's dissatisfaction after lamellar keratoplasty. A wide range of postoperative refractive error from −13.0 D to +7.0 D is reported after lamellar keratoplasty [64, 65]. Topographic astigmatism of greater than 4 D has been reported in 16–34.4% of the patients [64, 65].

There are several options to treat astigmatism following lamellar keratoplasty that vary from optical correction using glasses or rigid gas-permeable contact lenses to surgical interventions including relaxing incisions, femtosecond laser arcuate keratotomy, wedge resection, photorefractive keratectomy, laser in situ keratomileusis, and toric phakic intraocular lens implantation [66]. Spectacles may be insufficient when a significant amount of astigmatic anisometropia is present, and rigid gas-permeable contact lenses may be an option only if they are well tolerated by the patient [66]. Astigmatic keratotomies can be precisely performed by using a femtosecond laser. However, this technology is not widely available, and its cost-effectiveness needs to be taken into account [66]. Excimer laser photoablation techniques are capable of treating coexisting spherical refractive error, but their efficacy is limited in the correction of high degrees of astigmatism. Manual relaxing incision, performed on the steep meridian, is widely used for high astigmatism after keratoplasty because it is a safe and simple procedure with no risk of postoperative haze and minor manipulation to the allograft. This technique, however, has some disadvantages including unpredictable results and risk of corneal perforation [66]. Corneal perforation can occur during manual relaxing incision because it is difficult to evaluate precisely the depth of the blade by conventional en face microscopy. Microperforations that present a slight leak without the development of a shallow anterior chamber usually self-seal and require no further interventions. A bandage contact lens and systemic carbonic anhydrase inhibitor can be used to manage such microperforations. If a shallow anterior chamber develops, however, the site of perforation should be sutured using 10-0 nylon suture material.

3.2.3.2.4. Graft immune rejection

Although lamellar keratoplasty eliminates the risk of endothelial rejection, other types of graft rejection (epithelial and stromal) may still develop with an incidence between 3 and 14.3% [67, 68]. Epithelial and stromal graft rejections after lamellar keratoplasty are very similar to those following penetrating keratoplasty and can be reversed by frequent topical steroid [68]. Epithelial and stromal graft rejections after lamellar keratoplasty must be treated appropriately to prevent less severe but still important complications including graft vascularization and suture abscess which can result in graft opacification and even failure [69]. Each episode

of graft rejection is treated with 0.1% topical betamethasone every 1–4 hours, based on the severity of rejection. The topical corticosteroid dose is gradually tapered over 2 weeks after resolution of the rejection episode [21].

3.2.3.2.5. Graft opacification and interface vascularization

Recurrence of opacification (scars and deposits) can occur at two sites: the graft itself and donor-recipient interface. Recurrence of opacification and deposits in the graft are frequent observations after keratoplasty in mustard gas-induced corneal involvements as a consequence of dry eye, limbal ischemia, limbal stem cell deficiency, and the presence of leaking vessels [57]. The advantage of lamellar keratoplasty is that it can be repeated with ease when recurrent graft opacity precludes useful vision or causes ocular irritation.

Donor-recipient interface opacification is a unique complication following conventional lamellar keratoplasty that develops due to recipient bed roughness, inadequate tissue removal, inadequate donor-recipient adhesion, or postoperative vascular invasion. Surface and suture complications may stimulate vascularization of the graft and interface. Extensive vascularization may result in lipid and protein extravasations leading to interface opacification (**Figure 7**) and hence visual acuity reduction. A full-thickness graft is still inevitable when deep stromal scar or visually significant interface opacity develops [58].

3.2.3.2.6. Interface keratitis

The interface left during lamellar keratoplasty is a potential dead space and microorganisms inoculated intraoperatively have a chance to proliferate within this space away from recipient immune response. Candida species is the most common microorganism obtained

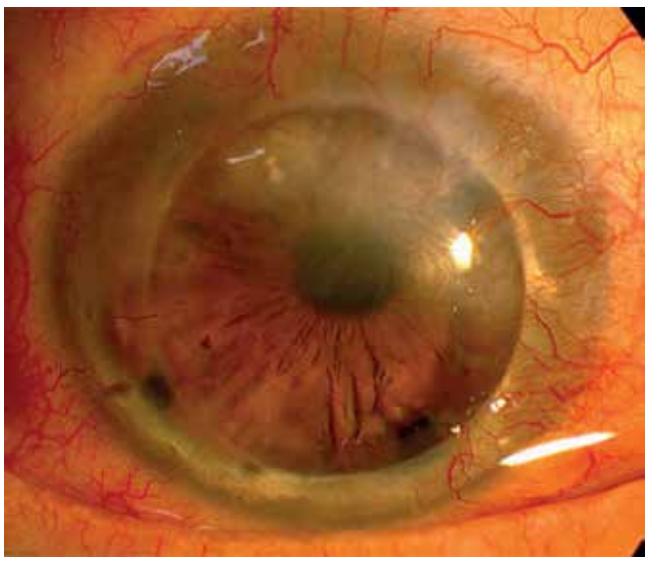

Figure 7. A lamellar corneal graft complicated with dense interface haziness and vascularization 4 years after transplantation in a mustard gas victim necessitating penetrating keratoplasty.

from interface keratitis [70]. Infection with this microorganism occurs because of donor corneal contamination or by the indigenous microflora of the conjunctiva and ocular adnexa. Recipient Descemet's membrane separates the site of infection from the intraocular structures after transplanting a contaminated donor cornea to the recipient bed. However, the location of infection may make it more difficult to obtain specimens for culture. Furthermore, it may prevent adequate penetration of topical, intraocular, and systemic antibiotics, making conservative treatment more likely to fail. Given that, both topical and systemic antibiotics should be prescribed, but penetrating keratoplasty may be needed to eradicate the infection [70].

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Chapter 2

Why Do Patients with Controlled Glaucoma Continue to Lose Their Vision?

Shimon Rumelt and Schachar Schreiber

Additional information is available at the end of the chapter

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Abstract

The question why patients with controlled glaucoma continue to lose their vision and may end with blindness was raised at the conference last year, but no answer was provided. This presentation will address some of the possible clinical causes such as supine position during sleep and sleeping on the affected eye(s). Antihypertensive drugs at bedtime increase the risk of anterior ischemic optic neuropathy, which is a challenge to diagnose in advanced glaucoma. Basic causes include the continuation of neuronal apoptosis despite controlled intraocular pressure. To prevent further visual loss in these patients, practical steps such as sleeping at 20–30° head-up position, avoiding sleeping on the affected eye(s), avoiding taking antihypertensive drugs at bedtime, and developing antiapoptotic drugs such as antibodies are essential.

Keywords: glaucoma, visual field damage, scotoma, blindness, prevention, intraocular pressure, supine position, antihypertensive drugs, apoptosis

1. Introduction

Glaucoma is a group of diseases that affect the optic disc, causing a specific type of optic neuropathy characterized by specific changes in the optic disc and visual field that eventually may progress to blindness. A feature common to most of the glaucoma types is high intraocular pressure, which to date is virtually the only target for treatment. The aim of the treatment is to decrease the intraocular pressure to a target pressure that is specific for each patient. This specific pressure is supposed to prevent further deterioration in visual field and irreversible blindness. Unfortunately, despite achieving a target pressure, there is a subpopulation of glaucoma patients who still progress gradually to blindness. This chapter will discuss possible reasons for this phenomenon in those patients who achieve the target pressure.

2. Epidemiology of blindness

The World Health Organization (WHO) estimated that in 2014, 285 million people (4%) out of the 7.2 billion world population had either low vision (246 million) or blindness (49 million) [1]. Ninety percent of these live in low-economic settings and 82% are aged over 50 years. Eighty percent of visual impairment can be prevented or cured. The best examples are correction of refractive errors and cataract surgery.

The most common cause for blindness worldwide is cataract (47%), and it is reversible upon surgery. The second common cause for blindness is glaucoma (12%), and it is the most common cause for irreversible blindness. This is followed by age-related macular degeneration (5%).

3. Blindness from glaucoma

Glaucoma is a distinctive group of optic nerve neuropathies characterized by specific optic disc and visual field changes, usually with an increase in intraocular pressure (IOP). In the past, a true IOP of up to 21 mmHg was considered normal in healthy individuals. Today, some consider an IOP between 18 and 22 as borderline. The term "true IOP" addresses the corrected IOP according to the thickness of the cornea and other parameters that influence the IOP. The main optic disc change is the increase of the cup (cupping) and decrease of the rim that contains the axons from the retinal ganglion layer (**Figure 1**). Early signs for this include disc notching, increased excavation, retinal nerve fiber defects, and papillary flame-shape hemorrhages. The early changes in visual field include Bjerrum defects (scotomata), paracentral scotoma, nasal step, and arcuate scotoma. As the disease progresses, visual field defects increase, first in depth and then in size. Arcuate scotoma may join nasal field, and when these increase toward the center and the periphery, tunnel vision and/or a temporal crescent or a

Figure 1. An advanced stage of glaucomatous optic disc damage showing thinning of the rim. The cup/disc ratio is almost 1 (subtotal excavation).

few visual islands remains. Eventually, these disappear too and the patient remains with no light perception. The changes in visual field correspond and follow closely with the changes in the optic disc. The chronic forms of this group are asymptomatic until advanced and irreversible visual loss occurs. Patients preserve normal visual acuity (even of 20/20) in one or both eyes until late in the disease. Such patients may not be aware of the small defects early in the course of the disease or even advanced concentric visual loss and tunnel vision, until they completely lose their vision in one or both eyes.

Two theories explain the neuronal loss in glaucoma. The first claims that mechanical force exerted on the optic disc causes direct destruction. The second claims that compromised blood flow causes damage. The damage may be caused also by a combination of these two processes. The end point is apoptosis of the ganglion cell layer.

4. Pattern of visual loss in glaucoma

The visual field loss (scotoma) in glaucoma has a distinctive pattern that differs from visual loss due to other causes (**Figures 2** and **3**). The visual field defects include Bjerrum scotoma, paracentral scotoma, nasal step, and arcuate defect. These defects correspond to retinal nerve

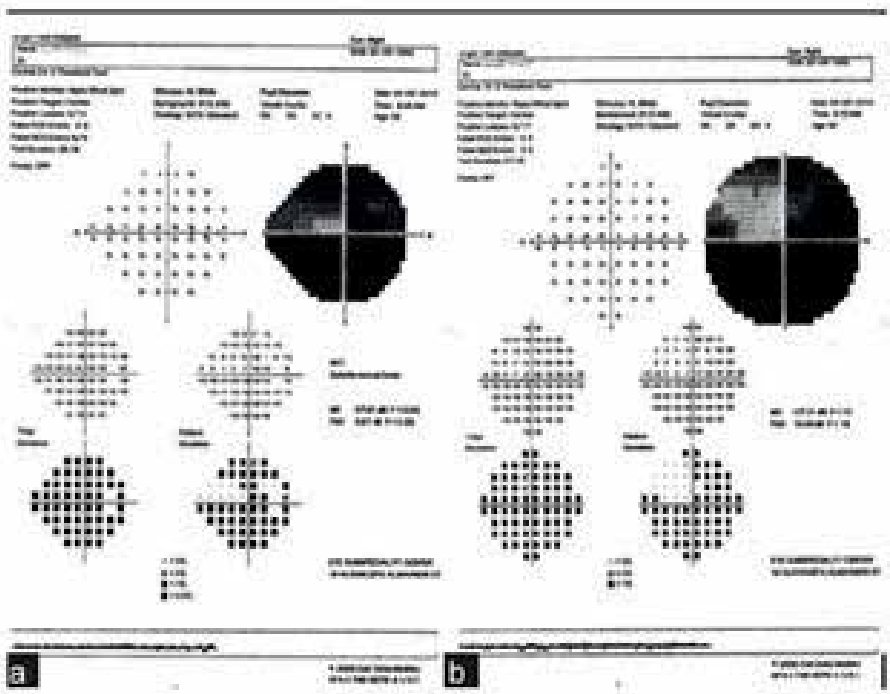

Figure 2. Advanced visual field loss of the right eye in a glaucoma patient. On the left, a 24-2 Humphrey visual field demonstrating a concentric visual field loss with only a small para-central island remained. The fixation point is split. This is also demonstrated in the same patient on the right with a 10-2 visual field. Glaucoma surgery at this point can cause the loss of the fixation and a decrease in best-corrected visual acuity to counting fingers.

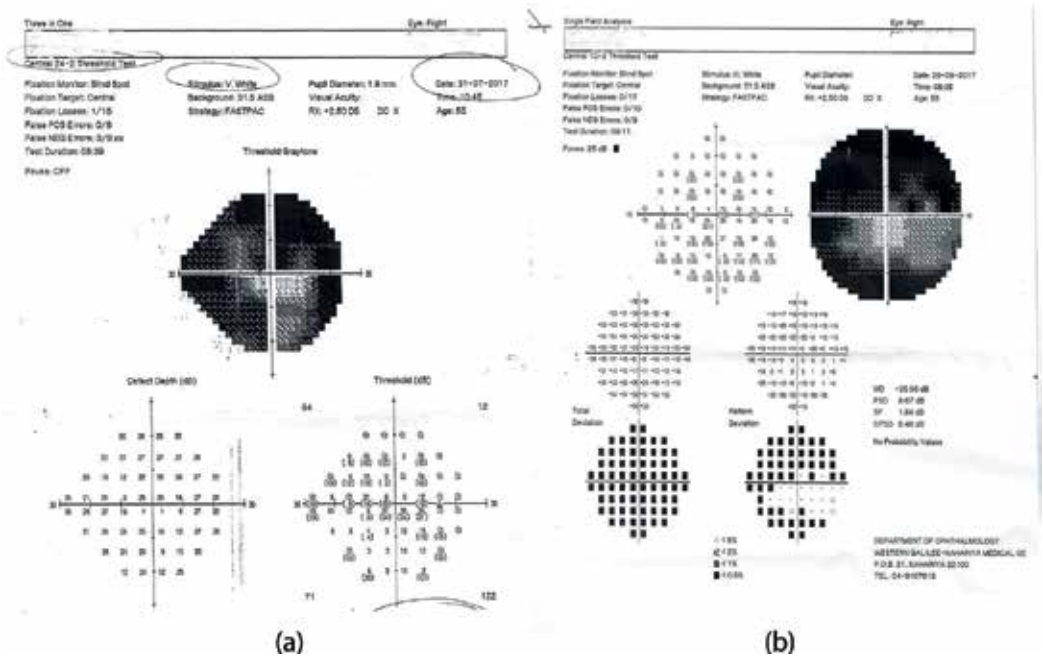

Figure 3. 24-2 and 10-2 Humphrey visual fields of a patient with advanced field damage. The damage encroaches on the fixation but does not split it. In this case, successful trabeculectomy with mitomycin-C was performed. The visual field remained unchanged. The best-corrected visual acuity remained 20/60, and the intraocular pressure decreased from 28 to 34 mmHg to 10–12 mmHg and remained at this level.

fiber loss, which usually begins in the arcuate bundles and the nasal fibers and ends with the papillomacular bundle.

When superior and inferior nasal steps coalescence with arcuate defects and spread centrally and peripherally, tunnel vision evolves. The visual acuity may remain intact (best-corrected visual acuity 20/20) in this situation. Eventually, central vision and/ or temporal peripheral island(s) may remain; when these are lost, the patient remains with no light perception. Occurrence in both eyes results in total blindness. In most types of glaucoma, the chronic ones, the patient may not be aware of the visual field loss, unless comparing each eye to the other. This is the reason that glaucoma is called the silent thief of vision. The patient may present only when the visual acuity in one eye is completely lost because the overlapping between the visual field of both eyes, micro-saccades, and most importantly, turning the head toward the area of interest. Therefore, screening of the population is the most crucial measure to detect glaucoma patients and treat them early.

The goal of treatment is to stabilize the visual field and prevent further deterioration in visual field and visual loss. Unfortunately, currently, the main treatment is aimed only at reducing the intraocular pressure (IOP) and achieving the ideal IOP (target IOP), which differs for each patient and is determined by the type of glaucoma, its severity, progression, patient compliance, and allergy to medications. In general, it should be as low as possible but not too low (hypotonia). Screening of the population includes observing the optic disc and checking

the IOP. It should be performed at least every 5 years before the age of 40 years and every 6 months after the age of 40 years.

5. Prevention of visual loss

To date, visual loss in glaucoma is irreversible because of the death of ganglion cells and their axons. The treatment is aimed to prevent continuous visual field loss and is divided into medical, laser, and surgical methods. To prevent visual field loss, the intraocular pressure should be at or below the target IOP, which is individual to each patient and related to the type of glaucoma, severity of the disease, patient's compliance, and allergy to medications. To date, there is no treatment addressing the different genetic defects and molecular mechanisms causing or related to glaucoma. The first line of treatment is usually medications. To enhance treatment, laser treatment may be applied. Some of the laser treatments such as selective laser trabeculoplasty have a short span of effectiveness, usually 1–1.5 years. If these fail, surgery is indicated. The number of medications, laser, and surgical procedures is wide and is determined mainly by the type of glaucoma.

Screening for glaucoma should include the entire population and should be composed of observation of the optic disc and documentation of the cup-disc ratio (C/D ratio) and other features of glaucomatous optic disc damage and intraocular pressure. Screening is usually performed every 5 years and over the age of 40 years twice a year. Patients with higher risk for glaucoma (e.g., family history of glaucoma, pseudoexfoliative syndrome, pigmentary dispersion syndrome, borderline IOP, etc.) may be routinely evaluated more often. Every patient who is diagnosed with glaucoma should be on appropriate medications permanently unless successful surgery has been performed, and even than the patient should be routinely followed.

The follow-up is every 3–4 months for lifetime including after successful surgery. If aggravation occurs, the follow-up intervals may be more frequent. Examination should be performed at different hours of the day, and a diurnal curve is indicated for patients with controlled IOP under medications and continuous visual field damage. The diurnal curve is performed every 4 hours and may even be increased to every 2 hours under medications. Some types of glaucoma such as pseudoexfoliative and pigmentary have a high fluctuation rate that may be missed by routine IOP examination. It is imperative to perform surgery in a timely manner, before the glaucoma is too advanced and before splitting of the fixation on visual field testing. Patients with complete splitting of the fixation are at higher risk to lose their central vision after surgery. Except for glaucoma surgery, other procedures may be required and may result in decrease of IOP. Cataract surgery in presence of risk factors such as hard nucleus (brown, red or black cataract), pseudoexfoliation, phakodonesis, lens subluxation, small pupil, ocular surface disorders such as ocular cicatricial pemphigoid, and Fuch's corneal dystrophy should be performed early. As the number of risk factors increases, surgery should be performed earlier. Visual field should also be obtained for these patients before surgery, if the glaucoma is advanced (C/D ratio of 0.9 or more).

Patients at high risk to lose their vision are those who do not take their medications regularly and/or do not follow-up with their ophthalmologist at regular intervals as indicated above.

Other major factors for visual loss are late diagnosis that may occur with all types of chronic glaucomas and slow decision making. Aggressive glaucoma and poor surgical outcomes may contribute to visual loss.

6. How to define controlled glaucoma?

The aim of treatment at present is controlling the IOP to prevent further deterioration in visual fields. The loss of visual field is irreversible. The ideal IOP should be low enough to prevent visual field loss without compromising the functions of the eye. Each patient has a desirable range of IOP—target IOP, which varies between individuals and depends on the aggressiveness of the disease. The aggressiveness of the disease is determined by the IOP, its fluctuations, the type of glaucoma, and the damage to the optic disc and visual field. In normal tension glaucoma, the target IOP is usually less than in other types of glaucoma, because even with normal pressures, the damage continues to progress. The IOP is constantly changing. It depends on the hour (diurnal variations) and seasons. Most but not all subjects have the highest peak in IOP during the early morning.

To be considered as "controlled glaucoma," the IOP should be within its target during the entire day in a long follow-up with constant use of anti-glaucoma medications or postoperatively. The patients should take their medications properly at a preset times and continuously. Thus, patients intolerant to anti-glaucoma medications or uncompliant are not considered controlled. The IOP may be assessed by diurnal curve every 4 hours, usually between 8 AM and 8 PM, because it changes constantly or even every 2 hours.

7. Matters of definition

In this chapter, controlled glaucoma was defined as target IOP under diurnal curve of 4 hours in patients, who are dedicated in taking their anti-glaucoma medications or after surgery. It is a philosophic question whether patients who continue to lose their vision are controlled. Perhaps the definition should be patients who do not show further signs of deterioration. However, since the target pressure has been achieved, it is expected that the patients will demonstrate stability of their visual functions (i.e., visual fields), and this may not occur in a subset of these patients.

8. Reasons for progressive visual field loss despite controlled glaucoma

8.1. High IOP fluctuations

Secondary glaucomas such as pseudoexfoliative and pigmentary glaucomas have high fluctuations of IOP, which varies depending on the dispersion of pseudoexfoliation material or pigment in the anterior chamber angle. The IOP peaks are unpredictable and variable in time and amplitude and may be missed by diurnal curve even if performed every 2 hours. They may occur between the IOP measurements and may be missed. To overcome this, frequent IOP monitoring including at bedtime and at shorter intervals may reveal such patients. Patients

with high and large fluctuations that are on full medical treatment may benefit from early glaucoma surgery, either trabeculectomy with mitomycin C or shunt procedure. Still, patients without IOP fluctuations may progress to blindness from other reasons as stated below.

8.2. Increased IOP in supine position (at bedtime)

People spend about one third of the day (6–8 hours) sleeping. The resting time may increase after retirement. The IOP increases at supine position compared with standing or sitting in healthy subjects by 2.47 ± 2.12 mmHg (mean ± standard deviation) ($p < 0.001$) when measured by non-contact tonometer Keeler, Pulsair EasyEye [2]. In another study, the IOP in sitting position was found to be 13.5 ± 2.0 mmHg in the right eye and 13.2 ± 2.3 mmHg in the left eye in healthy individuals [3]. The IOP increased in supine position to 16.8 ± 2.3 mmHg and 17.0 ± 2.3 mmHg, respectively ($p = 0.001$). This may result in deterioration of the optic disc and visual fields. Diurnal curve has probably no meaning if the patient is awakened at bedtime, and the pressure is measured while sitting.

The intracranial pressure (ICP) may also influence the progression of glaucoma [4, 5]. The ICP is directed through the subarachnoid space opposite to the IOP through the lamina cribrosa, and the difference between them is the translaminar pressure gradient. Theoretically, if this is low, the progression may be slower than if it is high but this may not be true. A high ICP and IOP with a low gradient may be sufficient to cause increased optic disc damage because of the increased shearing force in the lamina cribrosa and decrease in axonal plasma flow. This may initiate or facilitate axonal apoptosis.

8.3. Increased IOP when sleeping on the affected eye(s)

Most ophthalmologists do not live with their glaucoma patients and have no idea about their behavior in daily life. The patients may sleep on their affected eye(s), and this causes further increase of the IOP in addition to the increase caused by supine position. When the eye leans against the bed or pillow or when the entire mass of the head is over all or part of the globe, IOP is increased by 33%. Thus, the physician should inquire about the sleeping habits of the glaucoma patients. Actually, increase in IOP measurement can be seen in patients who squeeze their eyes during evaluation with Goldmann tonometer, as well as with some other instruments. It can also be seen if the examiner presses the globe during IOP measurement.

8.4. Antihypertensive drugs at bedtime

Glaucoma patients are usually older and have many associated aging and pathologic conditions, including atherosclerosis and systemic hypertension. Other ischemic diseases such as diabetes mellitus may also be encountered. Taking antihypertensive drugs before sleeping increases the risk for anterior ischemic optic neuropathy (AION) [6]. Antihypertensive medications decrease the perfusion into the optic disc, and this may join atherosclerotic changes in the blood vessels. AION may be difficult to diagnose in patients with advanced glaucoma. In advanced glaucoma, the cup may be large (cup/disc ratio of 0.8 or more), and the rim is thin enough not to distinguish pallor of the rim following additional AION. In addition, AION field defects may be superimposed on the glaucoma visual field defects. In advanced glaucoma, the visual field scotomata may be large enough (e.g., tubular vision) to prevent detection of the additional scotomata caused by AION. According to the vascular theory, damage to the optic nerve may be caused

also from ischemia if the optic disc does not receive enough oxygen even without AION. This damage is added to the damage caused by the mechanical effect of optic disc compression.

8.5. Continuation of the neuronal apoptosis

Patients with glaucoma suffer loss of axons of the ganglion cells as they pass the optic disc. Two theories explain the axonal loss. The first one is mechanical. According to this theory, the force caused by the IOP impedes axonal transport (flow) (micro-strangulation) and this may trigger axonal apoptosis [7]. The second theory is vascular. This means that the IOP impedes vascular supply to the optic disc. This causes a relative ischemia to the optic disc and triggers apoptosis. It is probable that both mechanisms coexist and the mechanical force may have a greater influence. Nonetheless, apoptosis, and not degeneration/necrosis, is the mechanism of axonal death in glaucoma. Apoptosis is programmed cell death, while necrosis is a different process involving extracellular components of inflammation. It consists of several pathways initiated be certain extracellular ligands such as programmed death ligand 1 (PD-L1), Fas ligand (FasL), tumor necrosis factor (TNF), nerve growth factor (NGF), growth factors, and others (**Figure 4**) [8, 9]. These molecules attach to receptors on the cell wall such as tropomyosin kinase receptor (TRK), tyrosine kinase receptor (RTK), receptor of apoptosis signal factor (Fas), and tissue necrosis factor receptor (TNFR) that trigger intracellular cascades that involve multiple pathways and molecules including the caspase cascade. These processes

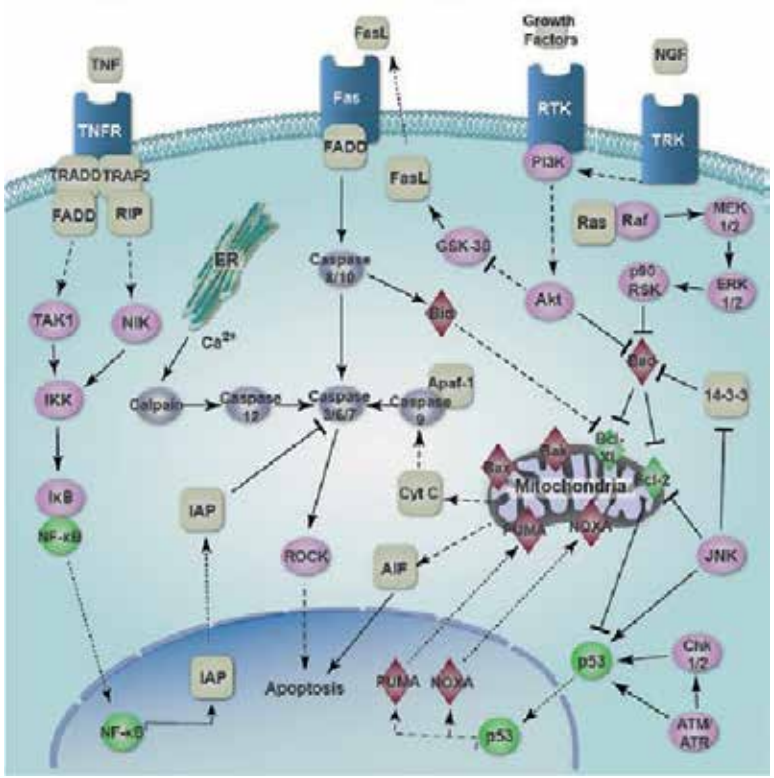

Figure 4. The pathways of apoptosis. Interference with any of these steps may prevent the apoptosis cascade.

occur in the cytoplasm, endoplasmic reticulum, and mitochondria that lead signals to the nucleus to degenerate. The end result is shrinkage of the nucleus, fragmentation of the deoxyribonucleic acid (DNA), and death of the cell. It is possible that some additional mechanisms and pathways exist that involve adjacent cells such as astrocytes, oligodendrocytes, and even vascular endothelial cells. Despite controlled IOP, the apoptosis may continue once started causing additional ganglion cell death. Ganglion cells in different stages of apoptosis may "signal" adjacent normal cells to commence with apoptosis cascade, leading to further loss of neuronal cells.

9. Recommendations to prevent further visual loss in patients with controlled glaucoma

Patients with high IOP fluctuations are not controlled and may benefit from early surgery such as trabeculectomy with mitomycin C or shunt procedures. These patients can be traced because they usually have secondary glaucoma mainly pseudoexfoliative and pigmentary. It is worthwhile to ask the patients to sleep at 20–30° head-up position. The IOP decreases when the bed head is tilted up in 30° and is 14.2 ± 2.3 mmHg OD and 14.1 ± 1.9 OS and not when the patient is sleeping on multiple pillows (16.3 ± 2.4 OD and 16.5 ± 2.6 OS) [3]. In another study, the IOP decreased from 16.02 ± 1.65 to 14.5 ± 1.36 mmHg [10]. The IOP may decrease by 9.33% in glaucoma patients, and this effect is found in 82% of them. Patients should avoid sleeping on their affected eye(s). Sleeping over the back or even on the side as long as the orbital rim is lying against the pillow is the best option for these patients. Antihypertensive medications should be taken when the patient is awake and active, usually in the morning and not at bedtime. It is the physician role to make these recommendations.

Additional efforts should be made to discover drugs that can abolish or slow down the apoptosis. Antibodies against PD-L1, FasL, growth factors, or their receptors may be helpful. Forty chemical compounds have inhibitory effects on different steps of apoptosis but may be toxic to normal cells. Phenoxodiol, an isoflavone that targets a regulator of sphingosine kinase depriving the cell of XIAP and FLIP was evaluated for ovarian cancer but was disappointing. Thus, it is essential to discover biologic agents such as antibodies against one or more of the extracellular mediators with better effects and with few or no side effects that will be approved for clinical use to arrest axonal apoptosis at the optic nerve. So far, none has been discovered, and research efforts are mandatory because such molecules may be used in glaucoma as well as other fields to prevent cellular few or no by apoptosis.

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Chapter 3

Visual Loss in Neuro-Ophthalmology

Eitan Z. Rath

Additional information is available at the end of the chapter

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Abstract

Optic neuropathy is damage to the optic nerve from any cause. Damage and death of these nerve cells lead to characteristic features of optic neuropathy. The main symptom is loss of vision (visual acuity and visual field damages), with colors appearing subtly washed out in the affected eye. The diagnosis is made on clinical examination. The history often points to the possible etiology of the optic neuropathy. In most of the cases, one eye is affected but it could be both. A rapid onset is typical of demyelinating, inflammatory, ischemic, and traumatic causes. A gradual course points to compressive, toxic/nutritional, and hereditary causes. The classic clinical signs of optic neuropathy are visual acuity and field defects, dyschromatopsia, and abnormal pupillary response. There are ancillary investigations that can support the diagnosis of optic neuropathy. Visual field testing, neuroimaging of the brain and orbit are essential in many optic neuropathies including demyelinating and compressive. In the last decade, increase of use new technology for optic neuropathies evaluation including multifocal visual evoked potentials and optic coherence tomography. Long standing of optic neuropathy is described by pale optic disk or optic atrophy, which means damage and death of these nerve cells or neurons.

Keywords: optic neuropathy, optic neuritis, non-arteritic anterior ischemic optic neuropathy (NAION), arteritic anterior ischemic optic neuropathy (AION), traumatic optic neuropathy

1. Introduction

Accurate medical history is very important information, helping to evaluate the etiology of visual loss. Rapid onset is characteristic of optic neuritis, ischemic optic neuropathy, inflammatory (non-demyelinating), and traumatic optic neuropathy. On the other hand, gradual onset over months or even years is typical of compressive toxic/nutritional optic neuropathy. A history over years is seen in compressive and hereditary optic neuropathies. The

ophthalmologist or neurologist can make the differential diagnosis according the symptoms, the age of onset, and the gender. Young age group (15–45 years) for optic neuritis women gender and pain on eye movement are more typical for optic neuritis versus. Elderly patients (older than 50 years), painless loss of vision without gender predisposition are typical for ischemic optic neuropathy. Additionally, in a young patient, history of neurological symptoms such as parenthesis, limb weakness, and ataxia is suggestive of demyelinating optic neuritis.

In an elderly patient (more than 60 years mostly 70–80 years) with signs of severe optic neuropathy and the presence of preceding transient visual loss, temporal pain, jaw claudication, fatigue, fever, anemia, weight loss and myalgia, an arteritic ischemic optic neuropathy (AION) due to giant cell arteritis (GCA) should be suspected.

In children, a history of recent flu-like illness or vaccination days or weeks before vision loss points to a para-infectious or postvaccinia optic neuritis, respectively.

Transient visual obscurations, transient diplopia, and headache should raise the suspicion of increased intra-cranial pressure.

The use of any medications should be carefully noted, since some are either directly or indirectly toxic to the optic nerve. These include drugs as ethambutol, methanol, isoniazid, tobacco alcohol, and more. History of diabetes mellitus, systemic hypertension, hypercholesterolemia, coagulation deficit, and smoking is more common in patients with nonarteritic ischemic optic neuropathy (NAION). Patients who have history of malignancy may have infiltrative or para-neoplastic optic neuropathy. It is important to inquire into the patient's general health, eating, and social habits (drinking and smoking) in suspected nutritional optic neuropathy (complex B–vitamins). In addition, a detailed family history is inquired in diagnosing hereditary autosomal and mitochondrial optic neuropathies.

This chapter addresses the major diseases neuropathies accompanied by rapid visual loss: nonarteritic and arteritic optic neuropathy, traumatic optic neuropathy, and optic neuritis.

2. Anterior ischemic optic neuropathy (AION)

Anterior ischemic optic neuropathy (AION) is a medical condition involving insufficient blood supply of the pial vessels originating from the choroidal vessels to the optic disk. AION is generally divided into two types: arteritic AION (or AAION) and nonarteritic AION (NAION) [1, 2].

We have to differentiate between two different etiologies, and therefore, workout prognosis and treatment possibilities are different.

2.1. Nonarteritic ischemic optic neuropathy (NAION)

NAION is the most common cause of sudden optic nerve-related vision loss. It is estimated that the incidence of NAION is about 8000/year in the USA and encountered for 90% of the optic neuropathies. NAION is mostly unilateral [3] and rare bilaterally. NAION is more

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frequent in Caucasians, no gender predisposition, and mean age at onset in most studies is from 57 to 65 years. No clinically effective treatment exists because little is known about its pathophysiology, and there are only few histopathological studies of the acute condition.

NAION [1, 2] typically presents suddenly upon awakening the painless patient notes seeing poorly in one eye. Vision in that eye is obscured by a dark shadow, often involving just the upper or lower half of vision. On examination, the patient is found with visual acuity reduction from 20/25 down to hand movement only, relative afferent pupillary defect (RAPD), swollen disk (segmental or diffuse) with splinter hemorrhages (see **Figure 1**), absent of large cup, and contralateral disk is small and crowded in 20–40% of the patents [4]. In approximately 6 months following the infarct visual acuity improves by 3 or more lines of vision on the Snellen chart in 42.7% of patients. In addition, vision had worsened by 3 lines or more in 12.4% of patients; some clinicians use the term "progressive ischemic optic neuropathy". Second eye involvement occurs in approximately 20% of patients with NAION within 5 years. Furthermore, most cases of NAION involve the loss of an altitudinal hemifield (**Figure 2**) (either the upper or mostly lower half of the visual field, but not both), and visual acuity remains almost normal or slightly reduced.

Figure 2 shows a few cases of NAION, which involve almost total loss of vision. The mechanism of injury for NAION is used to be controversial. Experts have come to a consensus that most cases involve two main risk factors. The first is a predisposition in the form of a type of optic disk shape named crowded disk [4, 5] or "disk at risk," where the cup/disk ratio is low (0.0–0.1), and secondly, cardiovascular risk factors as diabetes mellitus, hypertension, hypercholesterolemia, and coagulation deficits. Laboratory examinations at the presentation to differentiate between NAION and AAION include (erythrocyte sedimentation rate [ESR] that should be less than 40 mm/h) and C-reactive protein (CRP). It is advised to draw complete blood count and serum chemistry especially glucose, serum cholesterol and triglycerides, coagulophatic state, antitrombin III antiphospholipid antibody, and serum fibrinogen. Analysis of brain MRI suggests an increasing number of ischemic white matter lesions.

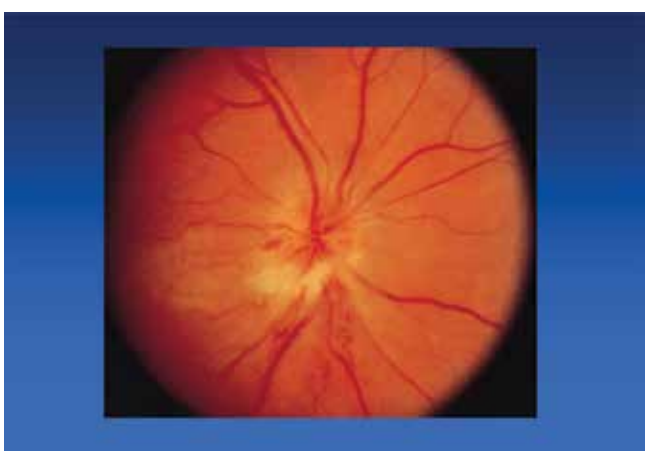

Figure 1. Disk appearance in nonarteritic ischemic optic neuropathy.

Additional risk factor such as obstructive sleep apnea, migraine, and hyperhomocysteinemia, smoking and optic disk drusen [6]. Ipsilateral carotid disease does not seem to be a risk factor for NAION. Association between cerebral and cardiac vascular disease seems to be very circumstantial. Drugs associated with NAION are amiodarone, phosphodiesterase-5 inhibitors such as sildenafil [7], and interferon-a.

Most experts throughout the world believe that there is no accepted treatment to reverse the damage. However, a recent large study by Hayreh has shown that if patients are treated with large doses of corticosteroid therapy during the early stages of NAION, in eyes with initial visual acuity of 20/70 or worse, seen within 2 weeks of onset, there was visual acuity improvement in 70% in the treated group compared to 41% in the untreated group [8]. Hayreh and Zimmerman performed a nonrandomized, open-label trial of systemic corticosteroids for acute NAION, and the untreated group had more vascular risk factors than the treated group, and therefore, this study was very criticized and not accepted by most neuro-ophthalmologists worldwide.

2.2. NAION treatments attempted with no specific success to improve vision

- Medical [9–11]
- Diphenylhydantoin
- Systemic: aspirin [12], corticosteroids [8]
- Intravitreal: anti-VEGF agents (e.g., Bevacizumab), (see more information)
- Erythropoietin/erythropoietin receptor agonists
- Surgical: optic nerve sheath fenestration, optic neurotomy (see more information)
- Hyperbaric oxygen [13]

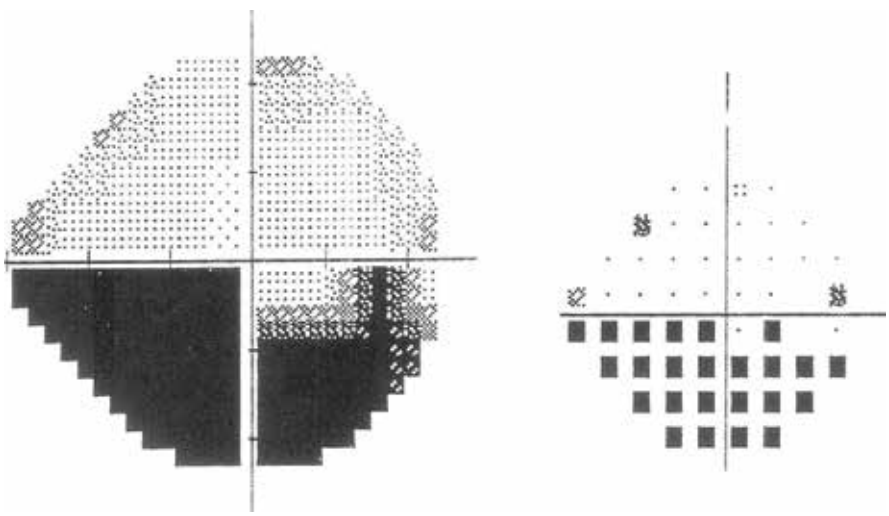

Figure 2. Visual field in NAION and lower altitudinal hemianopia.

2.2.1. Ischemic optic neuropathy decompression trial (IONDT)

The trial was done to assess the safety and efficacy of optic nerve decompression surgery compared with careful follow-up alone in patients with nonarteritic anterior ischemic optic neuropathy (NAION). The ischemic optic neuropathy decompression trial (IONDT) is a randomized, single-masked, multicenter trial and was carried out in 1994. Study was done by 244 patients with NAION and visual acuity of 20/64 or worse.

First group of 125 patients had been randomized to careful follow-up, and second group with 119 patients had been randomized to surgery, with 6 months of follow-up.

Patients in the surgery group received optic nerve decompression surgery and follow-up ophthalmologic examinations; those in the careful follow-up group received ophthalmologic examinations at the same times as the surgery group. A parameter of gain and loss of three or more lines of visual acuity on the New York Lighthouse chart at 6 months after randomization was used by the research group and measured by technicians.

Results showed that patients assigned to surgery did no better when compared with patients assigned to careful follow-up regarding improved visual acuity of three or more lines at 6 months: 32.6% of the surgery group improved compared with 42.7% of the careful follow-up group. According to the results, IONDT indicates that optic nerve decompression surgery for NAION is not effective and may be harmful [14].

2.2.2. Bevacizumab (Avastin®) trial

Intravitreal bevacizumab for the treatment of NAION neuropathy: a prospective trial [15].

2.2.2.1. Methods

In this non-randomized controlled clinical trial, 1.25-mg intravitreal Bevacizumab was compared with natural history [15]. Twenty-five patients were enrolled (17 with treatment and 8 controls). Patients were examined at baseline, 1, 3, and 6 months with a full neuro-ophthalmic exam, automated perimetry, and optic nerve optical coherence tomography (OCT) measurements. The primary outcome measure was change in mean deviation on Humphrey visual field testing. Secondary outcome measures were changed in visual acuity and optic nerve OCT thickness.

2.2.2.2. Results

(A) There was no significant effect of treatment on the primary outcome measure of mean deviation score ($P = 0.4$). (B) There was no effect of group assignment on the secondary outcome measures of change in mean Early Treatment Diabetic Retinopathy Study (ETDRS) letters ($P = 0.33$). (C) No change in nerve fiber layer thickness on OCT ($P = 0.11$).

Results show optic disc in NAION the results, there was no difference between Bevacizumab and natural history for change in visual acuity, visual field, or optic nerve OCT thickness [15].

2.3. New treatment opportunities (ongoing studies)

NAION is still an enigma regarding pathogenesis and treatment. The current therapeutically efforts are to preserve vision and minimize the damage from the primary insult. QPI-1007 is a small interfering ribonucleic acid (siRNA) designed to temporarily block cells from making Caspase 2 (controls cell apoptosis). Quark pharmaceuticals and NORDIC collaborated in a study that uses the possible effect of this drug as a possible neuroprotection therapy for NAION.

2.4. Arteritic anterior ischemic optic neuropathy (AAION)

Distinction between AAION and different etiologies of anterior ischemic optic neuropathy. will be discussed. AAION is due to temporal arteritis (also called giant cell arteritis (GCA)), an inflammatory disease of medium-sized blood vessels that occurs especially with advancing age (more than 60 years, mostly 70–80 years). Annual incidence rate in population age 50 years or older is estimated as 15–30/10,000. Female-to-male preponderance of 3.5:1 is prevalent in white population of European origin. GCA is associated with polymyalgia rheumatica. About 50% of the GCA patients have polymyalgia rheumatica, and 10–20% of the patients with polymyalgia rheumatica have GCA. Polymyalgia rheumatica may precede GCA or can occur simultaneously.

The symptoms and signs of severe optic neuropathy [16] include the presence of preceding transient visual loss, temporal pain, jaw claudications, fatigue, fever, anemia, weight loss, and myalgias are strongly suggestive of arteritic ischemic optic neuropathy (AAION). In contrast, NAION results from the coincidence of cardiovascular risk factors in a patient with "crowded" optic discs. Nonarteritic AION occurs in a slightly younger group and is much more common than AAION. Most cases of AAION involve nearly complete vision loss (light perception to no light perception), while only a few cases of NAION result in near total loss of vision (**Figure 3**). Swollen disk, elevated CRP, and ESR (60–120 mm/h) are highly suggestive of temporal arteritis (arteritic AION) [3, 17]. At times, the optic disk in AAION is characterized

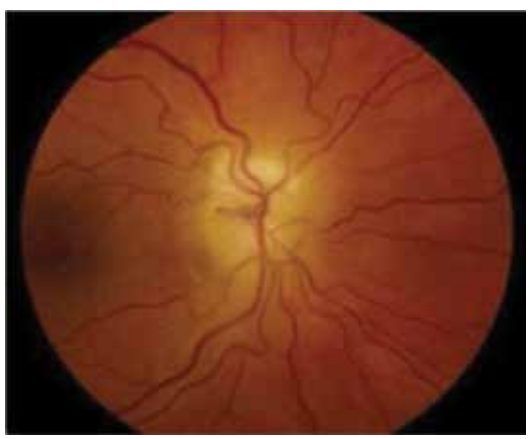

Figure 3. Shock white disk in arteritic anterior ischemic optic neuropathy.Ó 2018 American Academy of Ophthalmology.

by a milk-pale edema that may extend to the retina. In some cases, even central retinal occlusion with "cherry -red spot" may occur. Diagnosis is confirmed by temporal biopsy, and if the histological result is negative, it is necessary to biopsy the other side. Biopsy is taken from several segments along the temporal artery because the inflammation is segmental and may be missed by one site biopsy. Another possibility for diagnosis is ultrasound of the temporal artery but it is less accurate compared to biopsy.

Neuroimaging is not usually required in patients with typical presentations of giant cell arteritis (GCA) and when performance is generally normal [18]. However, some patients have already undergone imaging before neuro-ophthalmic evaluation, and these studies may be abnormal. They report four main imaging findings described in the literature:

1. Nonspecific orbital enhancement.
2. Optic nerve parenchymal enhancement.
3. Perineural sheath enhancement.
4. Optic chasmal enhancement.

Other important MRI findings in GCA include those involving the vascular supply not only extracranially but also intracranial, particularly vessel wall enhancement of the intramural ICA. MRI findings may hold some diagnostic value in distinguishing between A-AION as in GCA and in nonarteritic AION, in which they are typically normal. Differential diagnosis for these MRI findings can lead to inappropriate testing and delay diagnosis and treatment [18].

2.4.1. Summary of AAION

1. GCA is a vascular disorder that may result in devastating visual loss if not treated promptly.
2. Biopsy is the gold standard for diagnosing, and neuroimaging plays a role only in atypical presentations.
3. Neuroimaging findings in GCA are often nonspecific and can lead to delay in diagnosis and treatment.

Patients are hospitalized for evaluation and intravenous corticosteroid treatment with at least 1 g/day (3–5 days) of methylprednisolone followed by prednisone 1 g/kg for 10 days and then tapering down. The dosage is decreased to 20–40 mg/day in 3 weeks, and treatment is continued for 12–18 months. A steroid sparing agent is tocilizumab, a monoclonal humanized antibody against interleukin 6 receptor. The dosage is 1 g/day for 12–24 months, and it is not given in the first trimester of pregnancy. It can be combined with corticosteroids, and this allows decreasing the dosage of the later. During the follow-up period, the inflammatory parameters including sedimentation rate, CPR, platelets, etc. is monitored, and if they increase, the dosage is accordingly increased. Treatment is very urgent to avoid AAION in the fellow eye, as it can happen even within days or weeks after the first eye was affected. Treatment generally does not improve the vision of the affected eye.

3. Optic neuritis

Optic neuritis is a condition that produces abnormal vision loss without causing ocular abnormalities and we have to differentiate between typical and atypical optic neuritis.

1. **Typical optic neuritis:** Condition of visual loss caused by inflammatory demyelization of the optic nerve either idiopathic or associated with multiple sclerosis (MS). The myelin sheath is the target of attack.
2. **Atypical optic neuritis:** The nerve becomes inflamed as a part of uveitis or systemic inflammation treatment can help to improve vision.

3.1. Typical optic neuritis

Optic neuritis is a term used to refer to inflammation of the optic nerve, and it appears in two forms: (1) when associated with a swollen optic disk, it is called papillitis or anterior optic neuritis. (2) When the optic disk appears normal, the term retro bulbar optic neuritis is used. Acute optic neuritis is the most common type of optic neuritis that occurs throughout the world and is the most frequent cause of optic nerve dysfunction in young adults mostly women. In this chapter, we will provide information about the clinical profile of optic neuritis, its natural history, its relationship to multiple sclerosis (MS), and the efficacy of corticosteroid treatment according the Optic Neuritis Treatment Trial (ONTT) [19–22].

3.2. Demographics

The annual incidence of acute optic neuritis is estimated in population-based studies to be between 1 and 5 per 100,000 people in the general population [23]. The majority of patients with acute optic neuritis are aged between 18 and 46 years, with a mean age of 30–35 years. However, optic neuritis can occur at any age, and females are affected more commonly than males by a ratio of 3:1 to 4:1.

3.3. Clinical presentation

Clinically, there are three major symptoms in patients with acute optic neuritis: (A) central visual loss in 90% of the patients. (B) Pain especially is exacerbated by eye movement around the affected eye in more than 90% of patients. (C) Relative afferent pupillary defect (RAPD) in all patients with unilateral optic neuritis [24].

Loss of central visual acuity occurs within few hours to several days, and the degree of visual loss varies from very minimal reduction to counting fingers (in rare cases, complete blindness can be observed). The majority of patients describe central vision loss predominately, and some of them complain of peripheral field defects. The visual loss is monocular in most cases, but particularly in children, both eyes are simultaneously affected.

The presence of pain is a very helpful, differentiating optic neuritis from other causes of optic neuropathies such as anterior ischemic optic neuropathy, which produces painless visual loss.

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Examination of a patient with acute optic neuritis reveals evidence of optic nerve dysfunction [24]. In addition, color vision (especially red color) is typically impaired in almost all cases and is helpful to differentiate from other optic neuropathies.

A relative afferent pupillary defect (RAPD) is demonstrable with the swinging flashlight test in all unilateral cases of optic neuritis. Patients with optic neuritis may also have a reduced sensation of brightness and contrast sensitivity in the affected eye.

Visual field (VF) scotomas involve many forms of central or peripheral field disturbances such as ceco-central scotoma, inferior or superior altitudinal hemianopia, central scotoma, Bejerrum scotoma, hemianoptic defects, and more, almost any type of visual field defect (see **Figure 4**).

Presentation of optic disk in the acute phase is mostly normal with sharp disk margins and reddish color. Some of the patients with acute optic neuritis have minor degree of disk swelling with no correlation to visual acuity or visual field loss [25]. Over approximately 4–6 weeks, the optic disk in an eye with acute optic neuritis may become or remain normal or become pale, and most parameters of vision improve. In the chronic phase, the pallor of the optic disk may be diffuse or sectorial from my personal experience often the temporal part (42%) because the papillo-macular bundle is damaged in many patients with optic neuritis [26].

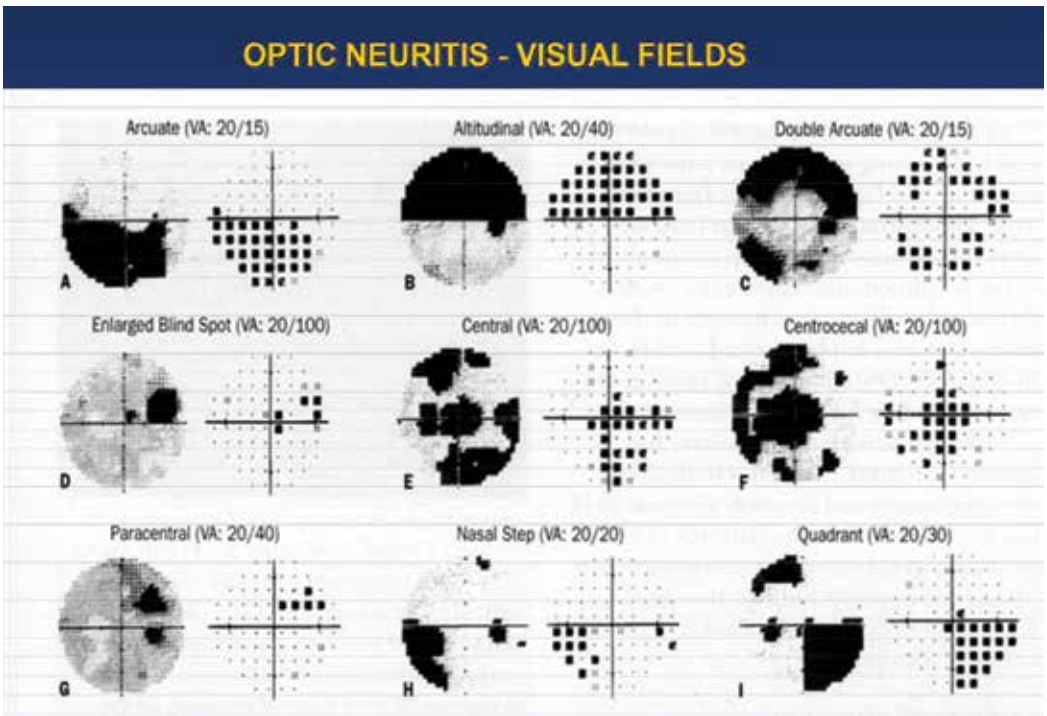

Figure 4. Visual field possibilities in optic neuritis.

3.4. Diagnostic studies

Imaging studies in patients with presumed acute optic neuritis are usually performed for the following reasons: (A) to rule out particularly a compressive lesion; (B) to determine if a cause other than demyelization is responsible for inflammation of the optic nerve; or (C) to determine the visual and neurologic prognosis of optic neuritis.

The best imaging study can be done by magnetic resonance imaging (MRI), it can reveal demyelization lesions of the optic nerve, manifesting as foci of T2-bright signal, areas of enhancement, and even optic nerve enlargement. These lesions are nonspecific, and a similar appearance can be observed in patients with infectious and other inflammatory optic neuropathies. The most important application of MRI in patients with optic neuritis is the identification of signal abnormalities in the white matter of the brain, usually in the periventricular region, consistent with demyelization. MRI is the strongest predictor of the eventual development of MS in patients with acute isolated optic neuritis. It can show multiple white-matter lesions in both cerebral hemispheres, including the periventricular regions.

Cerebrospinal fluid (CSF) analysis in the evaluation of patients with acute optic neuritis is not any more a strong predictor for MS. The presence of oligoclonal banding in the CSF is associated with the development of MS, but it can show false positive results. The powerful predictive value of brain MRI for MS is increased also because the Lumbar puncture examination is invasive. Therefore, CSF examination in the evaluation of patients with optic neuritis has been reduced. CSF studies in patients with optic neuritis are mostly useful to detect another inflammatory or infectious disorder.

3.5. Associated neurologic disorder

3.5.1. Risk factors for developing MS

The presence of at least 1 lesion in the periventricular white matter of the brain MRI is highly predictive, family history of MS, white race, old neurologic complains, winter onset, and younger age of optic neuritis. Conversely, patients with acute optic neuritis who have a normal brain MRI, severe disk swelling, a macular star, or disk hemorrhages or older age of onset have a low risk of developing MS.

The risk of developing MS [27–29] in a patient who experiences an attack of acute optic neuritis is about 75% in women and 34% in men over the subsequent 15–20 years, with the risk being greatest in the first 5 years after the first attack.

3.6. MRI diagnostic criteria for multiple sclerosis

Multiple sclerosis can be diagnosed when the MRI [30–32] in a patient with optic neuritis reveals two or more typical lesions of multiple sclerosis, at least one of which is contrast enhancing. The demyelization foci in the brain commonly appear in the corpus callosum and periventricular white matter and are best seen on T2-flair images.

The number of inactive typical white-matter lesions is the most important criterion for estimating the risk that the patient will develop multiple sclerosis [31]. Optic neuritis with two or

more noncontrast enhancing lesions typical of multiple sclerosis on MRI is called a "clinically isolated syndrome" and is associated with a high risk of MS. Multiple sclerosis arises in only 25% of patients in whom MRI reveals no foci of demyelination in the brain. If one or two such foci are initially present, the risk is 65%; if three or more are present, it is 78% [31].

3.7. Treatment

Many studies have shown that there are no data to support the efficacy in any treatment to alter the final visual outcome during a period of a year after optic neuritis. Treatment with corticosteroids is the main treatment option for patients with acute idiopathic optic neuritis. The prognosis for visual recovery after acute optic neuritis is very good also without treatment. According the ONTT (IV regimen of corticosteroids) [19–21, 32], steroid treatment should be delivered in acute optic neuritis if symptoms of 8 days duration or less. Begin with 3 days of intravenous Methylprednisolone in a dose of 1 g/day followed by 11 days course of oral prednisone at a dose of 1 mg/kg/day with a taper over 3 days. The ONTT was done in randomizing 457 patients with acute optic neuritis, comparing a group of patients following the IV steroid regimen versus a group of patients receiving placebo. The results of the trial showed that this regimen does not affect the final visual outcome of a patient, but it accelerates the recovery of vision compared with no treatment in the first 2 years. In addition, patients who experience an attack of acute optic neuritis should not be treated with low-dose oral prednisone alone because it provide no effect of visual outcome and may double the recurrence rate for optic neuritis.

The ONTT [19] results had another important aspect of treatment for acute optic neuritis regarding the possibility of having impact on the development of MS. Patients who were treated with the intravenous followed by oral corticosteroid regimen had a reduced rate of developing clinically definite MS during the first 2 years following treatment. MS developed in only 8% of patients who were treated according the corticosteroid regimen versus 17% of patients in the placebo group. This benefit of treatment was seen only in patients who had abnormal brain MRI at the time of onset of the optic neuritis. The protective effect was short and by 3 years after optic neuritis groups treated with ONTT IV regimen versus placebo groups had equal incidence to develop MS. These findings suggest that a patient with acute optic neuritis who has an abnormal brain MRI may benefit in the short term (2 years) from treatment with the IV/oral steroid regimen.

A number of agents other than or in addition to systemic corticosteroids have been found to reduce the risk of the development of MS following an attack of acute optic neuritis over a longer period of time than corticosteroids alone. The Controlled High-Risk Avonex MS [33] Prevention Study (CHAMPS), a randomized, double-blind, placebo-controlled trial that enrolled patients with a first demyelinating event, offer some help. Weekly intramuscular injection with 30 ug of beta interferon 1a (Avonex) to patients who had 2 or more white-matter lesions of at least 3 mm on a brain MRI together with a 14-day course of Methylprednisolone followed by prednisone lowered the probability for MS. The group of patients receiving interferon beta-1a had a 44% reduction in the 3-year risk of developing MS compared with those receiving placebo. In addition, patients in the interferon group had fewer new and enhancing brain MRI lesions.

To conclude, a clinician should discuss with a patient having acute optic neuritis the treatment benefits comparing to no treatment emphasizing that there is a good chance (more than 80%) that visual acuity will recover to 20/20 within a year without treatment. It is important to explain the patient the relation between optic neuritis and the chances of developing MS. No treatment affects the final outcome of visual acuity.

3.8. Visual prognosis

The natural history of acute idiopathic optic neuritis is to worsen over several days to 2 weeks and then to improve mostly rapidly. Improvement can continue to occur up to 1 year after the onset of visual symptoms. I had some patients of which improvement started only after 2 months but it is uncommon. The mean visual acuity 1 year after an attack of otherwise uncomplicated optic neuritis is 20/20, and less than 10% of patients have permanent visual acuity less than 20/40. Most parameters of visual function, including contrast sensitivity, color perception, and visual field, improve in conjunction with improvement in visual acuity. According to some investigators, most patients retain excellent vision for at least 15 years after their first attack [24].

Although the overall prognosis for visual acuity after an attack of acute optic neuritis is extremely good, some patients have persistent severe visual loss after a single episode. Furthermore, even patients with improvement in visual function to "normal" may complain of movement-induced photopias or transient loss of vision with overheating or exercise (Uhthoff symptom). The ONTT since 1992 has made it clear that the risk of a recurrence or a new attack is substantially higher in patients treated with low-dose oral prednisone as opposed to patients who receive no treatment or who are treated according the ONTT [19]. About 25% of patients who experience an attack of acute optic neuritis will experience a second attack in that eye or a new attack in the previously unaffected eye.

3.9. Atypical optic neuritis

Optic neuritis that develops before the age of 15 years or after the age of 50 years may be atypical. Many of these cases have no periocular pain, and visual decline is over few weeks. Atypical optic neuritis should be divided to three categories: infectious, immune, and Sarcoid. Most of them appear with disk edema.

3.10. Infectious optic neuritis

This may occur in meningitis/encephalitis [34] and is treatable. The pathogens could be bacteria (Homophiles, Streptococcus, Staphylococcus, spirochetes, or mycobacteria), protozoa as Toxoplasmosis, fungi as Cryptococcus or Aspergillus, or herpes viruses [35, 36]. Syphilitic optic neuritis can develop very rapidly from every stage of the disease. Tuberculosis causes meningitis. Lyme optic neuritis is rare and mostly associated with those who visited near New Haven, Conn, USA.

Another type of optic neuritis is called Leber's stellate neuro retinitis caused by *Bartonella henselae* responsible for cat-scratch disease. States with disk edema and within weeks, we can find star shape collection of hard micro-exudates (star-shape) at the macula called neuroretinitis [37].

3.11. Immune optic neuritis

Optic neuritis can appear within days or weeks after systemic influenza illness [34] or vaccination [38]; often binocular with good vision recovery [32]. Atypical optic neuritis is also associated with acute disseminated encephalomyelitis (ADEM), a condition in which multiple CNS manifestations occur at once; in most cases, patients recover and never recur. Some authors recommend high dose of corticosteroid treatment.

In optic neuritis, if an underling cause is found, it should be treated with either corticosteroid or immunosuppressive medications.

Optic neuritis is rare in Guillain-Barre syndrome, Crohn's disease, ulcerative colitis, behest's disease, Wegener's granulomatosis, and lupus erythematosus.

3.12. Sarcoidosis

When the optic nerve is involved, the vision declines and the optic disk might be swollen, with or without systemic signs. Vision recovers with corticosteroid therapy. Relapses are common [39].

- Recommended laboratory tests mostly for atypical cases
  - C-reactive protein
  - Complete blood count
  - Serum chemistry
  - Blood sugar
  - Vitamin B_{12}
  - Rheumatoid factor
  - Antinuclear antibodies
  - Anti-phospholipids antibodies
  - Anti-ds-DNA antibodies
  - Lupus anticoagulant
  - Serum angiotensin-converting enzyme test
  - *Borrelia* serology
  - Urinalysis
- Additional tests in case of "clinically possible differential diagnosis"
  - Anti-neutrophilic cytoplasmic antibodies (ANCA)
  - Extractable nuclear antibody (ENA) profile

- Auto antibodies against aquaporin-4
- HIV serology
- Human T-lymphotropic virus type 1 (HTLV-1) serology
- *Treponema pallidum* hemagglutination assay (TPHA), long-chained fatty acids
- *Mycoplasma* serology
- Urinary methylmalonate excretion

4. Anterior versus posterior traumatic optic neuropathy

Traumatic optic neuropathy is the name given to the syndrome of an optic neuropathy after head or ocular trauma in the absence of other causes [40]. Like any other optic neuropathy, there are variable degrees of visual acuity and visual field loss and an afferent pupillary defect if unilateral or significantly asymmetric.

Traumatic optic neuropathy is either anterior or posterior and within each category can either be direct or indirect. Trauma to the anterior optic nerve usually injures the central retinal artery and vein, which enter or exit the nerve approximately 10 mm posterior to the globe. This vascular injury often results in retinal infarct. Hemorrhages are usually the result of severing the pial vessels with or without disk edema and rarely manifestations of central retinal or branch artery occlusion, central retinal vein occlusion, or anterior ischemic optic neuropathy. Axonal injury in the posterior optic nerve does not cause any acute effects on the disk, nerve fiber layer, or retinal ganglion cell layers. Axonal transport abnormalities posteriorly do not affect the more anterior nerve fibers, and so disk edema is not seen in posterior traumatic optic neuropathy. For these reasons, isolated posterior traumatic optic neuropathy is associated with a normal fundus examination at presentation. Only after a few weeks, we can see the structural signs of optic neuropathy evident, namely disk pallor and thinning of the retinal nerve fiber layer. A particular type of posterior traumatic optic neuropathy is when there is injury to the chiasm, in which case, there may be unilateral or bilateral temporal visual field defects respecting the vertical meridian. Rare chiasmal injury can be seen with posterior avulsion of the optic nerve, for example, traumatic enucleation, or penetration from a foreign body.

4.1. Direct anterior traumatic optic neuropathy

Direct anterior traumatic optic neuropathy is defined when there is penetration of the optic nerve by a foreign body or projectile. Anterior direct optic nerve injuries result from medial penetrating orbital trauma that damages the anterior optic nerve, for example, a knife transecting the optic nerve just posterior to the globe. This is because the optic nerve course transverses the medical part of the deep orbit and is not protected there by the bones or the eye. Posterior direct optic nerve injuries result from penetrating orbital or head trauma more posteriorly, for example, a bullet that passes just anterior to the chiasm. Direct injuries tend to produce severe and immediate visual loss, with little likelihood of recovery. The reason

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for this presumably is that a major element in these injuries is transection injury to retinal ganglion cell axons, which causes instantaneous loss of axonal conduction and an inability to regenerate axons later.

4.2. Indirect anterior traumatic optic neuropathy

This is diagnosed when traumatic optic neuropathy occurs without a history of foreign body. It occurs in anterior indirect injuries, which associated with sudden rotation of the globe from blunt trauma. Examples include a digit trauma to the globe or falling and hitting the eye on the corner of a table. Anterior indirect traumatic optic neuropathy can cause partial or total avulsion of the optic nerve, with associated peripapillary hemorrhage.

4.3. Posterior indirect traumatic optic neuropathy

Posterior indirect injury is the most common cause of traumatic optic neuropathy. It results from blunt head trauma that transmits a concussive force to the optic nerve, resulting in contusion at the optic canal. There may be little or no evidence of significant head trauma; a fall from a bicycle may suffice. In other cases, there is multisystem trauma or significant brain injury. Loss of consciousness occurs in 40–72% of patients with traumatic optic neuropathy. Motor vehicle and bicycle accidents are the most frequent causes of traumatic optic neuropathy, accounting for 17–63% of cases. Traumatic optic neuropathy may be iatrogenic, especially after maxillofacial or endoscopic surgery as a result of inadvertent direct injury to the optic nerve or transmitted force fracturing the optic canal. The common site of posterior indirect optic nerve injury is at the optic canal; the intracranial optic nerve is the next most common site of injury. There may or may not be bone fractures. Despite being most common, posterior indirect traumatic optic neuropathies fortunately occasionally have the most favorable prognosis, its spontaneous visual recovery sometimes occurring at variable times after injury. Presumably, the injury causes concussion and focal blockade of axonal conduction without loss of its structural integrity. Once there is healing of the edema or other molecular events blocking conduction, axonal function can return. The severity of initial visual loss in patients with traumatic optic neuropathy varies from no light perception to 20/20, with sometimes only a visual field defect as functional evidence of disease. An afferent pupillary defect is always present and is the major clue for the diagnosis in the presence of otherwise normal eye. Patients with very poor vision (e.g., light perception only or no light perception) are less likely to improve, regardless of therapy, than patients with vision better than light perception. The reason is likely that severe injury causes axonal transection, membrane disruption, or cytoskeletal disorganization, any of which can lead to axonal dissolution and irreversible loss of conduction of visual information. In some cases, the visual loss only begins several hours to days after the injury. If this happens, the possibility of an intrasheath hemorrhage should be entertained, and neuroimaging should be repeated.

4.4. Neuroimaging

The diagnosis is radiological. It is essential in the evaluation of a patient with traumatic optic neuropathy not only for demonstrating correlative signs of injury but also detection of pre-existing

structural lesions and coincident intracranial effects of trauma, e.g., hematomas or carotid cavernous fistulas. CT scanning is superior to magnetic resonance imaging (MRI) in delineating fractures of bone. It is critical that CT be performed with very thin sections that are aimed to the optic canal, and reconstructions performed, particularly in the coronal plane. About 20 to 50% of patients with posterior traumatic optic neuropathy have evidence of an optic canal fracture by neuroimaging, and sometimes, the clue is a small loss of contour of bone. Although the displacement on neuroimaging may be small, it is possible that at the time of injury, there was a much larger displacement of the bone into the canal. Even in the absence of a fracture, blood in the sphenoid sinus should raise suspicion for optic nerve injury. MRI is better for imaging soft tissue, particularly the intracranial optic nerve and chiasm, and may be useful for delineating intrasheath hemorrhage that occurs at the orbital portion from penetrating injury (anterior direct TON). It is critical that MRI only be performed after a metallic intracranial, intraorbital, or intraocular foreign body has been ruled out by CT scanning or conventional radiography. If CT is used for screening, care should be taken to use thin slices and no interslice skip.

4.5. Treatment of traumatic optic neuropathy

In anterior and direct traumatic optic neuropathy, there is no evidence that treatment of anterior optic injuries or direct optic nerve injuries is efficacious. In the former, the concurrent vascular injuries cause direct ischemia and infarction to the neural retina and/or optic nerve head, and the time until irreversible neuronal death is measured in minutes to hours. In the latter, there is often sufficient direct axonal trauma to disrupt the integrity of the axon, up to and including its transection, and in the central nervous system of mammals, this is a point of no return for neuronal function. An exception is anterior traumatic optic neuropathy associated with neuroimaging evidence of an enlarged optic nerve sheath. In these cases, an optic nerve sheath fenestration should be performed in the hopes of evacuating an intrasheath hematoma.

4.6. Treatment of posterior indirect traumatic optic neuropathy

With respect to posterior indirect traumatic optic neuropathy, the three commonly used approaches that have been used are very high doses ("mega doses") of corticosteroids [41], decompression of the optic canal, and observation alone; there is insufficient evidence from good quality randomized trials to guide decision-making on how to treat traumatic optic neuropathy. Because visual function often spontaneously improves in this disease, clinical trials are particularly necessary for physicians to select therapies based on evidence. Megadose corticosteroids experimental models of white matter trauma in animals showed that doses of 15–30 milligrams per kilogram of intravenous methylprednisolone are protective for injured neurons [41]. The NASCIS 2 and 3 studies found that patients treated within 8 hours of spinal cord injury with a loading dose of 30 milligrams per kilogram of intravenous methylprednisolone load followed by 5.4 ml/kg/hr continuous infusion for 48 hours had a better outcome than control patients [42, 43]. Extrapolating these results to traumatic optic nerve injury, it was thought reasonable to believe that similar doses should be used for injury to this comparable central nervous system white matter structure. However, over the years, there has been controversy about interpretation of the NASCIS data [44, 45], and its application

to the treatment of spinal cord injury is not uniform [46, 47]. Furthermore, animal and cell culture data suggest that high doses of methylprednisolone may actually be toxic for the retinal ganglion cell and/or its axon [48–50]. Finally, the Corticosteroid Randomization After Significant Head Injury (CRASH) trial demonstrated that 48 hours of mega-dose methylprednisolone significantly increased the risk of death after head injury [51], with a hazard ratio at 6 months of 1.15 (95% CI 1.07–1.24) [52].

The authors concluded that "These final results still provide clear evidence that treatment with corticosteroids following head injury affords no material benefit."

4.7. Optic canal decompression

Decompression of the optic canal is usually achieved through the transethmoidal route, most commonly via an external ethmoidectomy or endonasally [53]. The canal is then decompressed inferomedially from the superior lateral wall of the sphenoid sinus, with care taken to avoid the carotid artery. Although the canal can also be decompressed through an intracranial approach, the former is less invasive. However, if surgery in the area is being performed for other reasons necessitating unroofing of the canal, then an argument can be made that decompression of the canal should be done through this approach. However, there is also no evidence that optic canal decompression is efficacious. A recent Cochrane review concluded that there is no conclusive evidence that any particular form of surgical decompression improves the visual outcome in TON. The decision to proceed with surgery in TON remains controversial and each case needs to be assessed on its own merits. The final decision will inevitably reflect a combination of clinical judgment, the availability of local surgical expertise, and the patient's perception of the possible risks and benefits. If surgery is to be considered, it should only be performed in centers with experience with the procedure. Because of the possibility that the carotid may be iatrogenic injured, there should be informed consent regarding the risk of death or stroke. Surgery should not be performed on an unconscious patient because of the difficulty in assessing visual function. Observation of traumatic optic neuropathy may improve without any treatment. There are no convincing randomized control trials to show a treatment benefit in traumatic optic neuropathy, and a nonrandomized concurrent comparative study did not demonstrate clear differences between treatments and observation. Therefore, when a patient cannot give informed consent for corticosteroid or surgical therapy, some neuro-ophthalmologists may simply observe the patient as none of these treatments have been proved to be superior.

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Section 2

Evaluation of Visual Impairment

Chapter 4

Glare and Ocular Diseases

My Diep and Pinakin Gunvant Davey

Additional information is available at the end of the chapter

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Abstract

Glare is the result of veiling luminance from the different light sources we are exposed to in our everyday lives. The luminance from glare can cause problems ranging from the discomfort of our eyes to vision loss. All individuals are affected by glare issues but those problems are intensified in patients living with ocular diseases. Therefore, understanding the effects of glare is applicable to elucidating visual function and pathology. This makes glare testing highly necessary in both clinic and research. However, there are many components involved in glare testing that makes attaining valid results difficult. This is evident in the flaws of current glare devices and the lack of a standardization of measuring glare. Despite the insufficiency of most glare devices, evaluating those weaknesses can potentially lead to a better understanding of glare and glare testing.

Keywords: glare, disability glare, cataract, glaucoma, macular degeneration, stereopsis, corneal diseases, keratoconus, glare testing, glare devices, contrast sensitivity, visual acuity, mesopic, photopic

1. Introduction

Our eyes are exposed to numerous light sources and at various intensities such as the rays from the sun or light from the headlights of driving cars. When we visually experience a veiling luminance from any light source it is a phenomenon known as glare. There are different types of glare: disability glare, discomfort glare, dazzling glare, and scotomatic glare [1]. We commonly experience discomfort glare when the intensity of the light source causes an uneasiness or annoyance on our eyes. Furthermore, we also regularly encounter disability glare. Disability glare is the scattering of light that enters our eyes that leads to visual impairment. Since disability glare directly affects our visual ability, it has been a focus of research, which particularly is important in an aging population and various disease states.

Light is focused to the retina to receive visual information of the world around us. Thus, the transmittance of light is integral to how we visually function. To this accord, the human visual system is finely tuned to allow the maximum amount of light transmission to the retina with least scatter. The retinal anatomy is also tuned to decreased sensitivity to shorter wavelength light and the retinal pigment epithelium and macular pigment allows the absorbance of stray light. However, disability glare interrupts the direction of light to the eye thereby interfering with the way we see [2]. This is especially debilitating, and the effects of glare are worsened in those who suffer from ocular pathologies. The many layers and components of the eye is involved in directing and processing light and cues to interpret our surrounding. Thus, a disease that impacts any part of the eye can exasperate disability glare decreasing the ability to see and perform daily activities such as driving.

The impact of disability glare makes it an important visual function to measure. However, currently there is no standardized way to measure glare [3]. There are both commercial and self-made device that hope to address this problem. However, more evaluation will be necessary to solidify their validity for research and clinical use. As a result, much of disability glare in visual function and pathology is still under research.

2. Pathological conditions

2.1. Corneal diseases

The major function of the cornea is to direct and refract light to the retina as well as provide structural support to the eyeball. Thus, preserving transparency and corneal shape is highly important in visual function [4, 5]. In various corneal diseases, the cornea is damaged through inflammation, swelling, and dystrophy [6]. The transparency of the cornea is the function of tight controls on water content, diameter of the collagen fibrils, and the spacing between the fibrils. The collagen fibrils have a diameter of 27–35 nm and the distances between fibrils are 41.4–60 nm [7]. The precise pattern of the collagen fibrils enables efficient light transmittance with minimal scattering or absorbance in a healthy eye. Any increase or decrease in the distance between the fibrils will compromise the transmittance of light [7].

Corneal edema is one example of a condition that disrupts the uniformity of these fibrils. The increased water content that results in edema changes the distance between fibrils, and thus can affect the overall transparency of the cornea. Reduced transparency, as we know, induces scattering when light enters the eye. Furthermore, scarring of cornea or deposits in the cornea can lead to the scattering of light as well. Post-surgical scarring is known to decrease vision and increase glare [8]. Additionally, certain medications like amiodarone causes cornea verticillata or deposits in the cornea that leads to the scattering of light rays [9].

Moreover, the type of light scatter that occurs can either be backwards or forward light scatter, depending on the angle of deviation light enters the eye. In backward light scatter, the scattering of light causes less light to reach the retina. While in forward light scatter, the scattering of light causes a luminance over the retinal image.

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Reduced transparency that leads to increased reflection and scatter of light can potentially cause disability glare. The disability glare along with diffraction and high-order aberration attribute to distorted retinal image, and thus impaired visual function. Components of vision such as contrast sensitivity can be hampered and if scattering is severe can lead to a deficit in visual acuity [4]. Therefore, those with corneal aberrations and abnormalities experience intensified forms of disability glare as well as reduced contrast sensitivity and visual acuity.

Keratoconus is a corneal dystrophy that leads to the progressive thinning of the center of cornea. Corneal thinning causes the center to protrude outward resulting in a cone shape cornea. Those with keratoconus can experience blurred vision as well as sensitivity to light [6]. Being reactive to light can make individuals with this corneal disease vulnerable to disability glare. Jinabhi and colleagues surveyed forward light scatter and visual function in subjects with mild to moderate keratoconus with no corneal scarring or history of ocular surgeries [10]. In the study, keratoconic and normal ocular healthy subjects underwent contrast sensitivity testing and glare testing to evaluate their visual function. The subjects with keratoconus exhibited lower contrast sensitivity than normal ocular subjects in testing. These results agreed with previous studies done and suggested contrast sensitivity was commonly compromised in keratoconus. Furthermore, keratoconic subjects also presented with intraocular scatter that resembled the increased scattering found in older populations or to those with early cataracts. Greater light scatter makes an individual with keratoconus more susceptible to disability glare [10]. More evidence of glare sensitivity in keratoconus could be found in a study done by Mäntyjärvi and Latinen. These researchers measured contrast sensitivity under glare conditions in keratoconic and ocular healthy subjects. The Pelli-Robson chart was used to measure contrast sensitivity. The chart contained letters of decreasing contrast that provided a quick and accessible way to measure contrast sensitivity [11]. The subjects were asked to read the Pelli-Robson chart under glare illuminance provided by the BAT. Then contrast sensitivity performance with and without glare was compared. The results of the comparison demonstrated that subjects with keratoconus experienced greater contrast sensitivity loss when tested under glare conditions than normal subjects [10]. Visual impairments being significantly greater in keratoconic subjects advocates the need for disability glare testing in measuring visual function. Disability glare performance can distinguish between normal individuals and those with ocular pathologies. Thus, in the case of corneal disease, disability glare can be a helpful diagnostic tool and could be a potential method of monitoring the disease progression. NEI VFQ (REF) or similar survey techniques can be used in conjunction to assist in evaluation of quality of vision and may be used in assessing glare related problems (**Figure 1**).

2.2. Glaucoma

Glaucoma is globally the second most common cause of blindness and it affects over – millions worldwide and is a very large socio-economic burden to the health care system [12]. The risk of glaucoma increases with increase age and elevated intraocular pressure is a major risk factor in glaucoma. Lowering intraocular pressure remains the only proven alterable risk factor that has shown to slow down the disease progression. Although, the exact pathogenesis in glaucoma remains to be identified, glaucoma leads to progressive damage to the to the optic nerve fiber

Figure 1. An optical coherence tomography image from a patient with early age-related macular degeneration. The drusen bodies are visible in the retinal pigment epithelium.

layer and changes in visual field that is in part associated to the level of intraocular pressure. If left unmanaged, glaucoma leads to progressive vision loss and blindness [12, 13].

Glaucoma affects several aspects of an individual's daily activities and task. Nelson and colleagues had articulated five major areas of difficulties in individuals with glaucoma. These difficulties include: (1) near vision issues, (2) peripheral vision issues, (3) dark adaptation and glare, (4) personal care and (5) household tasks, and outdoor mobility [14]. Their study measured both visual function and self-reported visual impairments. Subjects underwent multiple functional vision tests to assess the full spectrum of their visual capacity. The tests carried out included: Humphrey Visual Field Analyzer for visual field, Critical Flicker Frequency, Brightness Acuity Test (BAT) for disability glare, Goldmann-Weekers Dark Adaptometer for dark adaptation, Frisby Stereotest for stereopsis, and Farnsworth desaturated D-15 color test for color discrimination [14]. When comparing the results of the functional vision test to the self-reported impairments of the subject, there was a strong correlation between those two measures. Among the functional vision tests, disability glare testing done by the BAT best accounted for the difficulties the subjects reported. Nelson et al. also showed

that disability glare had one of the strongest relationship with the severity of visual field loss [14]. This relationship suggest that progression of glaucoma will be likely accompanied by increasing disability glare. Furthermore, the outcomes of this study affirm disability glare as a concerning visual impairment of glaucoma. In addition, the observed correlation between disability glare and visual field loss can potentially explain the components of the visual system that is involved in glare tolerance. This can in turn further the understanding of overall visual function.

As it is apparent that glaucoma patients suffer from disability glare, they found this impairment most concerning when driving. In surveying the value of various activities, glaucoma patients rate driving as highly important to maintaining their independence [15]. And so, understanding the impairments glaucoma patients face when driving is essential to addressing the concerns and preserving the quality of life for these individuals. Janz and colleagues surveyed open-angle glaucoma drivers and non-drivers about the types of visual problems they encounter during driving at a 6-month and a 54-month period. These surveys were also accompanied by ophthalmologic examinations. From the surveys and examinations, increasing visual field loss accounted for the differences between subjects who stayed drivers and subjects who became nondrivers because of their declining vision [15]. Thus, it can be inferred that those who are still drivers only had mild to moderate visual field loss. Despite little visual field loss, those drivers still reported many visual complications. One of the highest complaints from the drivers were tasks involving glare, which was said to be more troubling than visual search, peripheral vision, or visual processing speed which showed a lot of variation. Glare was a consistent issue among glaucoma drivers. Furthermore, glare was noted as one of the first issues subjects recognized when they first began to struggle with driving [15]. The study presents the driving challenges faced by glaucoma patients due to their sensitivity to disability glare. As mentioned earlier, driving is deemed as an important task to glaucoma patients to sustain autonomy. Therefore, assessing and managing disability glare is imperative to treating the visual impairments experienced by these individuals. Furthermore, since glare is one of the first detectable visual problems, disability glare test can potentially be utilized as a tool to identify the progression or worsening of a glaucoma in a patient. Though it is important to note that in the current state, it may be able to identify progression of the disease but may not give idea of the localization of the retinal damage in this disease. It will be interesting to evaluate the glare tolerance in various quadrants to see if the quantification of glare in specific locations is more sensitive than the non-specific glare tolerance testing.

2.3. Cataracts

The lens is a specialized structure that relies on its transparency, high refractive index, and curved surface to project clear images to the retina. Most of the lens comprises of concentric elongated fibers covered with an epithelium on its anterior surface. The epithelium along with the superficial fiber cells secrete an elastic extracellular matrix that encases the lens in what is known as the capsule [1]. Below the capsule, at the equator of the epithelium is where new fiber cells arise and differentiate [2]. The newer fiber cells constitute the periphery of the lens, named the cortex [1]. While the center of the lens is comprised of older fiber cells, some

originating from embryonic and fetal development, known as the nucleus [1]. Maintaining the transparency of the lens depends on the integrity of the arrangement of these fiber cells. However, as we age, oxidative damage and protein instability can accumulate, forming opacity in the lens and disrupting vision.

Cataracts is a disease cause by an opacification or cloudiness of the lens in the eye. The disease affects certain components of the lens, thus understanding the anatomy of the lens is important to pathophysiology of cataracts. There are various types of cataracts, but age-related cataract can be mainly divided into one of three types cortical, nuclear, and posterior capsular. Although, mixed type with features of three cataract types cortical, nuclear and posterior sub capsular are not uncommon. Each type has its own pathophysiology, anatomical differences and prevalence in the population [16]. Nuclear cataracts affect the oldest fiber cells of the lens which are the those formed in embryonic and fetal life. Evidence supports that nuclear cataracts arise due to the accumulation of reactive oxidative species that disrupt the normal protein and lipid components of fiber cells in the nucleus. The resulting cataracts causes patient to experience increase light scatter [17, 18]. However, cortical cataracts occur in matured fiber cells that arise later in life which lie closer to the surface of the lens. The progression of the cortical cataract encircles the outer circumference of the lens. The damages due to cortical cataract is much greater than that of nuclear cataract, the effects [17, 18]. On the other hand, posterior subscapular cataracts take place at the posterior surface of the lens where the cells just below the capsule are swollen. Since, the pathology of posterior subscapular cataracts is at the optical axis, visual function particularly reading tasks are greatly compromised. Furthermore, swelling of the posterior fiber cells impairs visual function even more by increasing the scattering of light [17, 18]. Clinically the cataract that causes the most glare related disability is the posterior subcapsular cataract. This is due two reasons (1) the entrance angle of the peripheral light rays is more oblique than central light rays and (2) the area that the posterior capsule cataract covers is also greater compared to nuclear cataract. Clinically in age related cataract we see mixed type of cataracts that has features that combine the nuclear, cortical and to some extent posterior subcapsular cataract.

The light is refracted through the lens before reaching the retina to be processed, and any sort of opacity that disrupts light transmittance can increase light scatter particularly if the opacity is large and spread throughout the lens. Being prone to disability glare, makes glare one of the biggest visual complaints and impairments experienced by those suffering from cataracts. Glare devices have an integral part in the research behind cataracts and currently, a large basis of literature is focused on the effects of disability glare on cataracts and how to accurately assess these visual challenges. Most glare devices available are geared toward cataract testing with the purpose of mimicking visual problems in real life in a clinical setting with the additional purpose of evaluating, monitoring and treatment of the disease state [2].

Clinically, cataracts are commonly evaluated by visual acuity charts which poses some problems. Visual acuity testing optotypes are at 100% contrast with black letters on white background and do not simulate real life scenario. In many cases, patients with cataract will have good visual acuities meeting legal standards of driving but still report experiencing visual

impairments while driving, difficulties in dimly lit environments and especially with disability glare [19]. Thus, the purpose of disability glare devices and testing methods is to provide additional information and insight that cannot be given with visual acuity testing.

There is evidence that supports that those with cataracts often experience a decrease in contrast sensitivity when compared to the age-match ocular healthy groups without cataract [20]. The contrast sensitivity loss in patients with cataract is even more pronounced under glare luminance [21]. Furthermore, cataract patients also have lower contrast sensitivity in mesopic conditions [1]. This becomes an issue when driving at night because that activity integrates mesopic light levels, contrast sensitivity, and the presence of glare. Thus, patients with cataract frequently complain of debilitating problems related to driving at night, under foggy, or rainy conditions, particularly with the addition of glare from incoming headlights [22]. Thus, as an importance of safety and the quality of life issue for those with cataracts, disability glare testing that accurately measures the challenges of night time driving is necessary. Disability glare in the daytime can also present visual impairments. Glare during the day predominantly originates from incoming rays of the sun. Unlike nighttime glare, daylight glare can be more accurately measured under photopic conditions [23].

There are numerous devices available that intend to simulate glare under the various conditions such as night, foggy, or rainy conditions, however, glare devices are not yet standardized [2]. Thus, the foundation on how to measure disability on those with cataracts have not been set. Though, the present literature already provided some insight to the impairments of cataracts. Research continues to find a valid, repeatable, and reproducible method for testing the disability glare.

Overall it is shown that glare induces a significant loss in visual function and individuals with cataract have further decline in visual acuity and contrast sensitivity in a range conditions with glare.

2.4. Macular degeneration

Centered at the retina is the macula which is highly packed with cone photoreceptors, and xanthophyll pigments that give it a darken appearance [24]. The macula is responsible for the majority of our photopic visual acuity, despite only comprising of less than 4% of the retinal space [24]. A disease known as age-related macular degeneration causes a gradual breakdown of these photoreceptors in the macula as well as changes in the retinal pigment. These damages lead to a decline in central vision [24]. Age-related macular degeneration (AMD) is divided into non-exudative (dry AMD) and exudative type (wet AMD). Early stages of dry AMD symptoms may go unnoticed, but patients slowly experience vision loss and can ultimately be converted to the wet AMD [25]. Some of the symptoms of AMD includes: decrease vision, blurry vision, metamorphopsia, and central scotomas [25].

As mentioned previously, the macula is comprised of xanthophyll pigments, specifically lutein and zeaxanthin. The role of these pigments is thought to have protective effects on the macula, as this is an area vital to visual function. Lutein and zeaxanthin are believed to filter some of

the harmful short-wave length blue light [24, 25]. Additionally, these pigments can also act as antioxidants to tackle free radicals and eradicate reactive oxygen species that damage the photoreceptors of the macula. Furthermore, lutein and zeaxanthin has shown to absorb straylight which can decrease the amount of harmful light entering the retina and possibly lower glare. The protective properties of these pigments led researchers to believe that increasing these pigmentations could potentially improve visual function.

One of the visual functions believed to be improved is disability glare and glare recovery. Stringham and Hammond looked at the relationship between disability glare and macular pigments. They measured macular pigment optical density (MPOD) of their subjects and compared that to their disability glare scores. The disability glare score was attained by measuring the level of illuminance from Maxwellian-View optical system that is high enough to induce disability glare when viewing sinusoidal gratings at 100% contrast [26]. From this test, the disability glare scores calculated showed a strong correlation to the macular pigment density. The researchers attributed the lower disability glare when there is a greater pigment density to the filtering effect of macular pigments. This was supported by the lack of correlation they showed between disability glare scores and macular pigment density when the glare source excluded the wavelengths of light that macular pigments are believed to filter [26]. These results provide compelling evidence for the involvement of the macula in disability glare. Disability glare is most associated with issues involving the optical media of the eye like the cornea and lens. However, as research has shown, the effects of disability glare can also be mediated by macular pigment. This provides more insight to visual function as well as the visual impairments that result from ocular diseases.

In additional studies, Stringham and Hammond recruited normal subjects who were given daily a 500-mg tablet that contained 10 mg of lutein and 2 mg of zeaxanthin over a 6 months period [27]. The research recruited 40 participants consisting of 23 women and 17 men. The subjects were assessed at 1,2,4 and 6-month period where their disability glare, photostress recovery, and macular pigment optical density (MPOD) were measured. As the researchers had done previously, disability glare was tested by utilizing the by the Maxwellian-view optical system to determine the illuminance level sufficient to cause visual impairment. All the subjects except for two had shown increase MPOD at the end of 6 months. The study subject also displayed reduced disability glare compared to baseline, tolerating greater veiling lights before any effects to their vision. On average, the participants tolerated 58% more glare ($p < 0.0001$) [27]. These results proposed a correlation between MPOD and tolerance to disability glare. This was further supported by the two subjects who did not experience any changes. These subjects that did not show an improvement in the MPOD also did not show an increased tolerance to glare. The researchers inferred that the macular pigment reduce glare disability by acting similarly to a yellow filter that cuts out short wavelength light and decreases veiling luminance [27].

From the relationship between macular pigment and disability glare, we speculate that the disability glare experienced by those suffering with macular degeneration can be partially due to the reducing level of MPOD. Moreover, knowing that MPOD can be supplemented and increased leaves possibility to improve the visual function of those with AMD, especially in the visual impairment of disability glare.

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3. Allied visual functions

3.1. Issues involved in glare testing

Disability glare plays an impairing role in many ocular pathologies such as the ones previously mentioned [2]. Thus, glare testing is not only valuable to understanding visual function, but it can also serve as a tool to evaluate the efficacy of treatments and surgeries of ocular diseases as well.

Though obvious that disability glare affects visual function, it still under study of what component of vision is most impaired by glare. Vision involves visual acuity, contrast sensitivity, stereopsis and many other components that can potentially be impaired by glare. Disability glare is commonly evaluated by either visual acuity or contrast sensitivity [28] (**Figure 2**). However, disability glare has shown to influence those aspects of vision differently, and so are important factors to consider when testing glare. Furthermore, glare is also tested under various light conditions such as photopic and mesopic. This is to mimic the changing luminance from day to night. Disability glare effects also varies from different light conditions; thus, presenting its own specific challenges in each light level [29]. Since glare testing is highly specific, appropriate variables must be incorporated for reliable and interpretable results.

Knowing the role of glare in visual function, proper glare testing methodology and devices are important. There are many components involved in glare testing some of which are the type of stimuli, glare source, and conditions. These factors play a role in the effectiveness of measuring disability glare and creating a real-world simulation. The capability of a glare testing method or device depends mainly on three criteria: discriminative ability, reliability, and validity [28]. Since glare methods and devices vary on the components they incorporate, so do their performance on the criteria mentioned. However, most current devices do fail to meet all three criteria, and thus there is still no standard way to measure glare. While there is a lack of standardization, there are a number commercial machines that are utilized in clinics and research [28]. Some of these devices are potentially valuable assessment tools but further research is necessary to evaluate their validity. However, there are many self-made devices created by researchers to address the glare test problem. Those have also shown good discriminative and repeatability. Despite positive findings, these devices and methods are still new and require much more additional research to assess their accuracy and validity.

3.1.1. Stereopsis

Stereopsis is the visual function of depth perception in a 3D world. The visual system integrates binocular disparity to interpret the placement of objects in space. Primarily a binocular visual function, good and balanced acuity of both eyes are necessary for proper depth perception [30].

As with some visual functions, stereopsis has shown to decrease with age even when visual acuity is still good. It is speculated that the decline in stereopsis is due to changes the eye undergoes with aging. The refractive and ocular motor system that can change with age can

Figure 2. Brightness acuity test (BAT) commonly utilized as a glare source for glare testing. Elliot et al. [28].

also influence stereopsis [30]. Alongside other visual functions such as contrast sensitivity and mesopic vision, disability glare has also shown to worsen with age [31]. Seeing a potential link, researchers considered the relationship between disability glare and stereopsis and whether they can predict the performance of one another. Schneck and colleagues measured coarse stereopsis and several other visual functions including disability glare in a population of individuals older than 58 years of age [31]. Disability glare was measured using a low contrast vision chart and a glare source. Further details of the disability glare testing were not given. These visual function tests were then analyzed on its relation to stereopsis. The results demonstrated that those who exhibited good visual function which included performing well on the

disability glare test were also those with good stereopsis. Similarly, when the visual function was low, their stereopsis performance was significantly lowered as well [32]. However, since the research grouped disability glare with other visual components in the analysis, there is no convincing evidence of a direct relationship with stereopsis. The inference that can be made is that an individual with healthy visual function should have both stereopsis and tolerance to disability glare intact.

Despite some correlational evidence, current literature does not show a strong relationship between stereopsis and disability glare. Though they are commonly assessed in visual function, stereopsis may not provide further insight to the effects of disability glare. Thus, glare testing seldom utilizes stereopsis as a measurement of visual performance.

3.1.2. Visual acuity versus contrast sensitivity

Glare testing consists of evaluating visual function under glare conditions. The most commonly used basis to determine visual function is contrast sensitivity and visual acuity [28] (**Figure 3**). The information provided by visual acuity and contrast sensitivity are utilized to determine severity of pathology, the need for ocular surgeries, and evaluate treatments. However, both these measurements convey different information, and so it becomes necessary to assess the validity of visual acuity and contrast sensitivity in evaluating glare. Furthermore, understanding how these measurements influence glare testing, it can provide us with further insight to what glare devices and testing techniques will ensue the most credible results.

Visual acuity is a familiar assessment done clinically using a chart with high contrast letters such as in the Snellen Chart or using symbols such as the Landolt C. The patient is asked to read the row of letters in assorted sizes at a set distance [33]. The smallest optotype the patient can read corresponds to their visual acuity [34]. Visual acuity has been shown to be a valuable tool to correct refractive errors. However, visual acuity has not been as effective in assessing target identification and detection [35]. Furthermore, the black letters on a white background found in visual acuity charts are not representative of the type of objects and conditions that are observed in day to day life. This is where visual acuity falls short of accurately portraying the visual difficulties one can experience in reality.

A less prevalent clinical evaluation is contrast sensitivity where varying levels of contrast is presented in the form of sinusoidal gratings, symbols, or letters. Much of contrast testing is done using sinusoidal gratings which has various phases, frequency, and contrast. The spatial frequency of the gratings correlates with sizes of realistic objects encountered in everyday settings. Low spatial frequencies have larger gratings, therefore is analogous to viewing larger objects. While higher spatial frequencies have smaller gratings, and thus analogous to viewing smaller objects [35]. Testing for contrast evaluates the various brightness and shades of gray commonly observed in real life.

Visual acuity and contrast sensitivity can provide overlapping visual information. Measuring visual function using high contrast and small letters in visual acuity is comparable to high contrast and high frequency optotypes in contrast sensitivity. However, contrast sensitivity has the advantage of incorporating a range of spatial frequencies, specifically low spatial

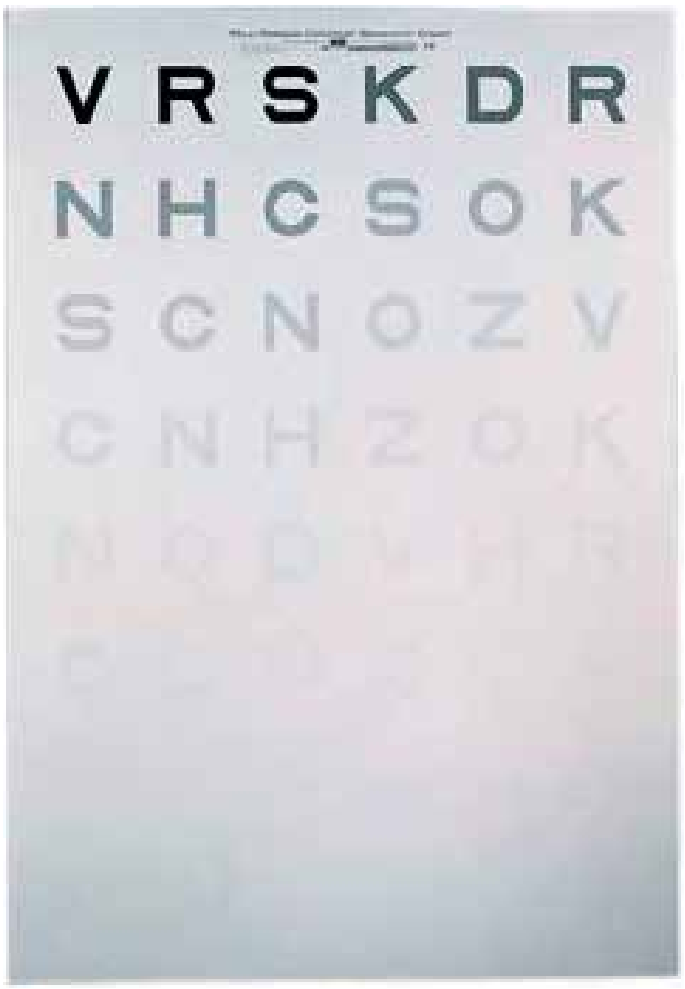

Figure 3. The Pelli-Robson chart tests varying levels of contrast but only at low spatial frequency. Image courtesy of Clement Clarke International Ltd. Elliot et al. [28].

frequencies which visual acuity lacks. Additionally, contrast sensitivity also supplies information on low contrast sensitivity which is often vision involved with nighttime [35].

There has been debate as to which measurement more accurately pertains to disability glare in real life situations. Increasing evidence in literature has shown that contrast sensitivity is a better predictor and more discriminative of disability glare in those with ocular pathologies than visual acuity. Those with cataracts often complain of visual impairments but measurements of their visual acuities meet normal standards. Hence, visual acuity may not be sufficient to identify problems caused by glare. Additionally, valuable information on visual function can be extracted by contrast sensitivity testing. A comparison study done by Elliot et al. looked at both the predictability of visual acuity and contrast sensitivity in subjects with early cataracts [36]. Since contrast sensitivity comprises of multiple factors, contrast was measured at high and low spatial frequencies. LogMAR charts with different contrasts was

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used to the measure the contrast sensitivity. In an age-matched evaluation of normal and cataract subjects, high spatial frequency contrast sensitivity showed the most visual impairments in subjects with early cataracts than low spatial frequency and visual acuity. An example of contrast sensitivity testing in low spatial frequency is the use of the Pelli-Robson chart. As this study has shown, low spatial frequency does not provide additional information or have good discriminative ability. This is further supported by another study completed by Elliot and his colleague, Bullimore. In their study, the Pelli-Robson chart in conjunction with the glare source from the BAT also showed poor discriminative ability. The researchers also believed this was attributed to the low spatial frequency of the Pelli-Robson chart [28, 36].

Furthermore, Abrahamsson et al. carried out a studied that assessed the sensitivity of visual acuity and contrast sensitivity to reflecting pathological differences under glare testing. Abrahamsson et al. was introducing a new methodology and device to test for glare [21]. The device had a point light source and used sinusoidal gratings as a measure of contrast sensitivity. The study used a glare score to analyze visual function between subject groups. The glare score was determined by using the lowest contrast visible to the subject. Once calculated, cataract and normal age-matched subjects were compared. Additionally, their visual acuity was also tested separately. By using contrast sensitivity as the basis of visual function, the glare device attained a disability glare score that correlated with the opacity of the lens in cataract patients. However, visual acuity showed a low correlation with the disability glare score, indicating that visual acuity may not be sensitive enough to detect changes in opacity [21]. These results suggest that contrast sensitivity tests can reflect subtle physiological changes. This can be beneficial to monitoring the progression of a disease and allow intervention before late stages. Also, contrast sensitivity can potentially lead to earlier detection of ocular pathologies.

While discrimination is necessary in glare testing, reliability is also highly important in attaining meaningful results. In the study done by Abrahamsson mentioned previously, the reliability of their glare device which used contrast sensitivity was good [21]. However, keep in mind that their retest was done on a small number of subjects and so further testing is necessary. While visual acuity tests have shown little discriminative ability, Elliot and Bullimore found glare testing that used visual acuity displayed high reliability. This is a potential positive in utilizing visual acuity in glare testing. The Berkeley Glare Test and the Regan charts using the BAT (Brightness Acuity Test) as the glare source are examples of glare tests using visual acuity [28]. In that same study, the evaluation of glare devices, Vistech and Miller-Nadler Glare Tester, which utilized contrast sensitivity demonstrated low reliability [28]. However, both those devices also exhibited little discriminative ability. Hence, the problem may reside in the design of the device and less so on contrast sensitivity. Moreover, the reliability of both visual acuity and contrast sensitivity is still not clear and their reliability needs to be further examined to determine its effectiveness in evaluating glare.

3.1.3. Lighting conditions

The measurements of visual function for disability glare are important considerations. However, it is also necessary to keep in mind that both visual acuity and contrast sensitivity perform differently depending on lighting conditions. Thus, one must consider the luminance

levels used during disability glare testing and how that relays to realistic encounters in everyday situations.

Contrast sensitivity performance in photopic conditions do not always correlate with mesopic conditions. Hertenstein et al. compared contrast sensitivity under both photopic and mesopic conditions [37]. Individuals recruited for the research comprised of normal, cataract patients, and glaucoma patients. The study utilized a glare testing device known as the Mesotest for the mesopic condition while using two different visual acuity test, Freiburg Acuity and Contrast Sensitivity Test (FrACT) and the Mars Letter Contrast Sensitivity Test for the photopic condition. Furthermore, the three testing methods were also retested to assure the reliability of the results. Overall, the study demonstrated that high mesopic contrast sensitivity score correlated with high photopic contrast sensitivity score. That correlation was also true when the subjects had low photopic contrast sensitivity score and low mesopic scores. However, high photopic contrast sensitivity score did not show the same predictability because individuals with those scores had various mesopic contrast sensitivity scores [37]. This suggest that to fully understand the visual impairments of disability glare, glare must be tested in different light conditions. Disability glare is present in everyday life at various light settings and so testing in many conditions provides more applicable knowledge of impairments patients face daily. As research has shown, visual performance differs depending on lighting and one condition cannot completely predict the results of another. However, testing under mesopic conditions may provide more information about visual function because a high score correlated to good vision in both light levels.

In patients with ocular pathologies and older drivers, concerns associated with disability glare often comes from difficulty driving at night. Realistic visual problems cannot always be accurately tested in clinical examination because visual acuity only tests visual function with high contrast and in photopics conditions. A study done by Kimlin and colleagues demonstrates this flaw by assessing the predictability of visual tests on the driving performance of its subjects [38]. These subjects had little to no ocular pathologies but had trouble night time driving. The subjects were put through driving obstacles to monitor their driving performance during night time. The subjects were also tested under photopic conditions for both visual acuity and contrast sensitivity. Then, they were tested under mesopic conditions for visual acuity and contrast sensitivity as well as glare testing. The study revealed that out of all the test results, high contrast visual acuity provided the least information about driving performance. In turn, glare and mesopic conditions were better predictors and accounted for more of the driving variations in the subjects [38]. Thus, a major visual problem like night time driving cannot be captured by typical clinical settings. Visual acuity and photopic conditions cannot provide information adequate in assessing all visual complaints. Thus, proper measurements of disability glare should be done in a lighting condition that most accurately addresses the visual complaint of interest.

In addition, mesopic conditions mimic those of night time illuminance as well as fog. While it has been shown that visual acuity decreases during mesopic conditions, central vision is less important and the ability to discriminate contrast becomes more necessary [39]. Thus, the effects of disability glare on contrast sensitivity during mesopic conditions can be more clinically valuable and applicable to daily life.

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4. Instruments and tests for glare

4.1. CSV-1000E

One widely known clinical tool to measure disability glare is the CSV-1000E from Vector Vision. This device measures disability glare using contrast sensitivity at spatial frequencies ranging from low to high. The spatial frequencies are measured using sinusoidal gratings at varying levels of contrast. The CSV-1000E has a backlit illumination of 85 cd/m^2 which can be used for glare testing under photopic conditions. The device can measure in mesopic conditions as well with the use of neutral density filters which lowers illuminance to 3 cd/m^2, the FDA recommended setting for mesopic measurements [40] (**Figure 4**).

The test consists of eight levels of contrast for each spatial frequency. There are eight columns consisting of two circles each, one which contains the sinusoidal gratings. The subject is tasked with identifying which of the two circles contain the grating for each of the columns. The responses are recorded and converted to a logarithmic scale.

Since the CSV-1000E can test in both photopic and mesopic conditions at various spatial frequencies, it has a variety of useful applications in a clinical setting. Shandiz et al. demonstrated the use of the CSV-1000E in individuals with different types of cataracts and different levels of severity. The CSV-1000E was sensitive enough to display a correlation between the subject's performance on contrast sensitivity and their level of lens opacity [41]. Since the CSV-1000E is a discriminative test that reflect ocular pathologies, it can be valuable in tracking the progression of a disease such as cataracts.

While the CSV-1000E has shown some discriminative ability, one report has shown the device is unreliable. Kelly et al. looked at the repeatability of the CSV-1000E in children and adults. The results indicated that the CSV-1000E has poor reliability. The reliability only improved in

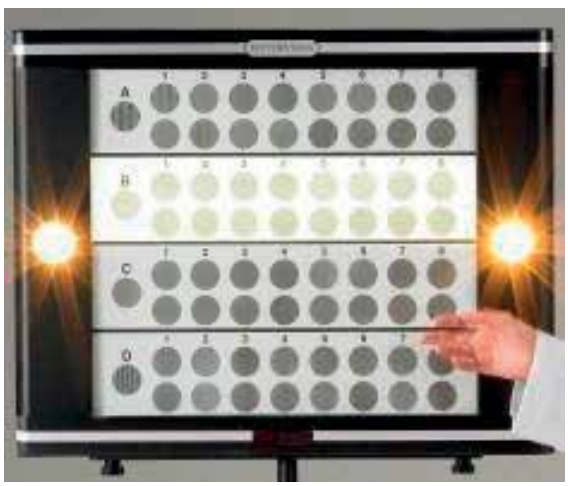

Figure 4. CSV-1000E used for glare testing at varying luminance and contrast sensitivity at different spatial frequencies. Image courtesy of VectorVision [40].

the case of maintaining the same experimenter, but even so the reliability was still low [42]. Some issues with the study was it involved both children and adults and the groups were too small to perform a reliable sub-analysis.

Examining the reliability of the CSV-1000E with a subject of pool of glaucoma patients, the investigators found the device and testing to be reliable. The reliability was calculated as the coefficient of repeatability (COR) which was on average .191 which was lower when compared to another known glare test, the Miller-Nadler Glare Tester (COR = 0.36) [43]. The study tested the effectiveness of a beta-blocker therapy on the contrast sensitivity of open angle glaucoma and looked at the reliability of CSV-1000E. The CSV-1000E was able to detect the changes in visual function from the beta-blocker treatment which can suggest good discriminative sensitivity [43]. Furthermore, based on repeatability the results supported that CSV-1000E can be a clinically reliable tool.

The CSV-1000E is a clinically versatile device as it can measure disability glare in various conditions. The device has also shown discriminative ability in detecting the changes in state of those with cataracts and glaucoma. However, the repeatability of the test remains uncertain and so further assessment of the CSV-1000E with a large sample size will be necessary for understanding its suitability in glare testing.

4.2. Halometer

Disability glare while causing a veil of light over the visual object, can also create an illuminated ring in our viewpoint which is known as a halo. The halo can be quantified by its disk radius and be used as a mean to measure disability glare. In a study conducted by Palomo-Alvarez et al., it was demonstrated that in comparison to straylight and corrected visual distance acuity (CVDA), disk halo radius was more discriminatively sensitive at detecting differences between normal and cataract subjects under glare conditions [44]. Thus, disk halo radius can be a valuable diagnostic tool to measure disability glare in clinics. One of the current tools for measuring halos are halometers. There are several models of halometers which are adopted by researchers to fit their studies. However, the foundational principals of the different halometers for evaluating disability glare are very similar.

The halometer test mainly entails a point light source at the center of the testing screen which varies in intensity depending on the device and study. The optotype used can be illuminated with a green or red light to monitor the effects of wavelength on light scattering. The protocol usually comprises of the subject moving the optotype either away or to the light source until it is just no longer visible or just visible depending on the specific instructions. The distant from the light to the object is then measured and analyzed as the disk radius halo which correlates with the amount of disability glare experienced.

In a study performed by Babizhayev et al., the halometer was used to assess individuals with cataracts [45] (**Figure 5**). Additionally, the performance of the Halometer was compared to other clinical tools such as visual acuity measurements and digitized opacity representations of the lens to determine the validity of the test. The digitized representations were done with retro-illumination photography that was digitally analyzed for light scattering and absorption.

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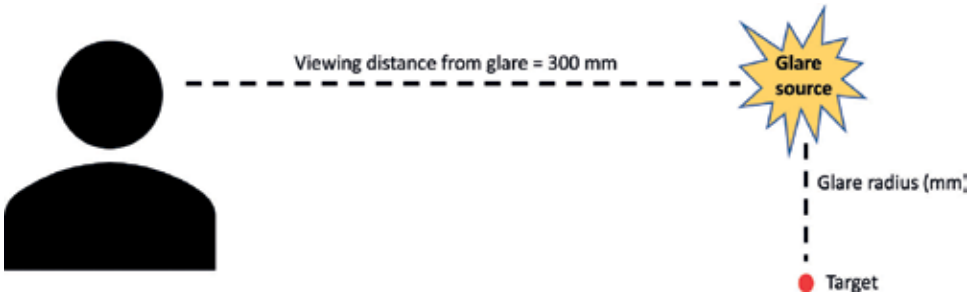

Figure 5. Schematic of the Halometer glare device utilized by Babizhayev and colleagues to measure intraocular light scatter in subjects with cataracts. Babizhayev et al. [45].

The halometer showed significant correlation between the visual acuity and the digitized opacity measurements. The results indicate that this glare test can contribute additional knowledge to visual function in relation to cataracts. Furthermore, the repeatability of the halometer was also assessed. The halometer performed with high repeatability of about 0.998 with test and retest occurring 1 week apart [45]. The halometer being both discriminative and reliable can be a beneficial and useful addition to clinical evaluation of patients.

Another modification of the halometer utilized an iPad application and an LED point light source. The halometer is known as the Aston Halometer [46] (**Figure 6**). The study subjects were tested monocularly with the use the Bangerter occlusion foil to induce disability glare. The target, presented at four different Weber contrast levels, was moved from the LED light source in eight different directions. The subject was to identify when the target was just visible from the light source and the distance, being the halo disk radius, was measured and analyzed. The performance of the Halometer was compared to the straylight meter which had been shown to be an accurate measurement of straylight and correlated to the amount of disability glare. The Halometer showed sensitivity to lower contrast letter and had high repeatability during testing which makes for a promising device [46]. However, the device was only tested on normal subjects without ocular pathologies. Therefore, while there is evidence in the Halometer's sensitivity to varying levels of contrast in normal subjects, the study did not provide insight to glare in ocular pathologies such cataract and glaucoma. Since the population of those living with ocular pathologies struggle with disability glare, a glare device needs to demonstrate discriminative ability in disease such as cataracts, glaucoma, and corneal disease.

Another study also used the Aston halometer to measure disability. They did so to evaluate night time driving in older adults with minimal pathologies including cataracts, glaucoma, and corneal pathology [38]. The subjects recruited was put through a driving obstacle to monitor their driving performance. Then mesopic conditions as well as glare testing was measured to see whether the visual testing is an accurate predictor of the subject's driving. While the test showed that the Aston halometer was a better predictor than photopic high contrast visual acuity (HCVA) testing, it was not a better predictor than mesopic high contrast visual acuity testing [38]. This suggest that the Aston halometer may need other improvements to increase sensitivity and further studies will be necessary to assess the validity of the halometer.

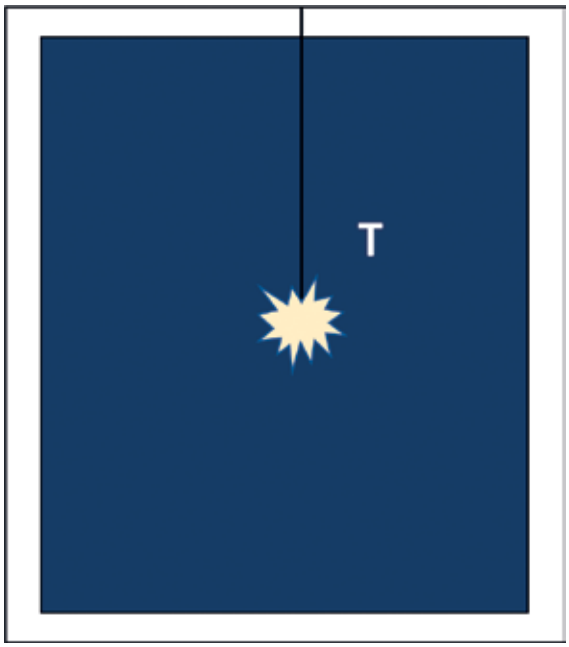

Figure 6. Schematic of the Aston Halometer designed on the iPad with an LED light source and target optotype controlled by iPhone. Buckhurst et al. [46].

4.3. Berkeley glare test

The Berkeley Glare Test has long been used for glare testing in clinic and research. The Berkeley Glare test measures visual acuity optotypes and different contrast levels under glare conditions [47]. A chart of varying levels of contrast is placed in the device, behind the opal Plexiglas screen which has a back illumination of 85 cd/m^2. The device has three levels of glare, being 300, 800, and 3000 cd/m^2. The creators of the Berkeley Glare test, Bailey and Bullimore, tested the technique on young and older ocular healthy adults [47]. Older adults were categorized as healthy if no ocular pathologies were present and their nuclear sclerosis score was grade 1 and under. The subjects were tested under four conditions which were no glare, and the three glare illuminations mentioned earlier. The chart used in the Berkeley Glare Test can vary and be chosen to meet specific needs. Visual function at high contrast was measured using the Bailey-Lovie Chart, a letter chart that assessed visual acuity. Low contrast visual function was also measured by using a letter chart that was at a Michaelson 10% contrast. The subjects were scored on a basis of a disability glare index (DGI) which was the difference in the number of letters the subject can see in the no glare versus glare conditions. Bailey and Bullimore's testing results showed that subjects with early nuclear sclerosis had a higher reduction in disability glare in comparison to visual acuity. The data also reflected subtle changes in lens opacity in the subject's DGI score before those changes could be detected by visual acuity testing [47]. The significant difference between DGI scores suggested that the Berkeley Glare test was more sensitive to physiological changes when assessing for contrast sensitivity than visual acuity [47]. This also noted the importance of using contrast sensitivity

over visual acuity in the case of the Berkeley Glare Test to produce more sensitive and accurate results. Furthermore, the Berkeley Glare Test also presented versatility as a glare device because the charts can be changed to test a wider range of visual function. This is potentially helpful in ocular diseases such as cataracts to evaluate different visual impairments in various settings. The Berkeley Glare Test also presented good discriminative ability as it can differentiate between those with early signs of nuclear sclerosis and normal subjects.

Further evaluation of the validity of the Berkeley Glare Test was done by Elliot and colleagues in a study where different glare tests were also observed [28]. The test was utilized with a low contrast (Weber 15%) Bailey-Lovie chart with a back illumination of 80 cd/m^2 and the glare setting was set to 750 cd/m^2 illumination. The Berkeley Glare Test displayed good repeatability but did not perform as well as the Regan chart and BAT (Brightness Acuity Test) as the glare source in reliability. The Berkeley Glare Test also exhibited good discriminative ability between normal and cataract patients. However, the study did disclaim that the subjects were referred to the ophthalmologist's office due to discrepancies in visual acuity. Since visual acuity in these subjects were already low, it can be expected that visual impairments were apparent enough to be easily detected by most tests. And so, these results did not further support the discriminative ability of the Berkeley Glare Test. The Berkeley Glare Test also fulfilled the three criteria of a vision test outlined by the American Academy of Ophthalmology (AAO). The criteria include: a force-choice protocol, test target follows a uniform logarithmic progression, multiple trials should be done at each level of acuity or contrast [28]. The Berkeley Glare Test's performance as outlined by the AAO criteria is both reliable and discriminative test. Therefore, the Berkeley Glare test can potentially be a strong foundation as both a research and clinical tool.

In another instance, a research study utilized the Berkley Glare Test to evaluate nighttime driving and disability glare. The study compared the Berkeley Glare Test to the Aston Glare Test in predicting night time driving performance. The Berkeley Glare Test did not show any significant correlation in driving performance while the Aston Glare Test displayed significant correlations [38]. This may suggest that while the Berkeley Glare Test can produce valid results, newer glare devices are surpassing it in sensitivity and leaves room for improvement in the test itself.

4.4. EpiGlare tester

In another glare test, known as the EpiGlare tester, the inventors developed a glare testing device that has the validity and discriminative disability to detect vision loss caused by glare. Epitropoulos and colleagues assessed the changes in corrected distance visual acuity (CDVA) in cataract and normal subjects under glare conditions [48]. The EpiGlare tester is a LED light emitting device that can be attached to a phoropter. There are four LED lights placed evenly around the aperture of the device. Under induced glare conditions, the subjects are asked to read off an EDTRS chart to assess their CDVA. The study also incorporated a Functional Vision Questionnaire that assessed the subjects driving and glare experiences. An additional question was asked after glare testing on how closely the test resembled their glare problems while nighttime driving (**Figure 7**).

Figure 7. EpiGlare tester designed by Dr. Alice Epitropoulos can be easily attached to phoropter for clinical use. Image courtesy of good-Lite. Epitropoulos et al. [48].

From the data of 40 subjects with cataracts and 49 ocular healthy subjects, EpiGlare tester demonstrated that cataract subjects are more impaired by disability glare than normal subjects [43]. These findings support the discriminative ability of the EpiGlare tester to distinguish the visual loss between pathology and healthy vision. Furthermore, the questions asked during the testing provides additional evidence to the validity of the device. From all the subjects, 83% of the cataract subjects reported the device accurately simulated their difficulties nighttime driving [48]. The device was easy to utilize and incorporate in clinical settings. The attachment to phoropter increases repeatability of the glare tester because the device setup will be consistent. The study did not directly examine its reliability and thus further evaluation of the device is still necessary. However, the EpiGlare tester simple use can be advantageous in clinical settings with its discriminative sensitivity and convenience.

4.5. Ophthimus glare tester versus contrast sensitivity function glare test

While there can be many variations among glare devices, the core of what is required in glare testing is the same. Therefore, there are several present methods and devices that share similar set ups. Two of which are the Ophthimus Glare Tester (Hightech Vision) and the contrast sensitivity function (CSF) glare tester created by Abrahamsson and his colleagues [21, 49]. Both these models examine cataract and normal subjects as well as monitoring their visual performance with contrast sensitivity under glare conditions (**Figures 8** and **9**).

These devices employed similar setups by using a ring fluorescent tube as the glare source with the optotype presented in the middle. Both assessed contrast sensitivity; however, the Ophthimus Glare Test utilized the Landolt C with varying levels of contrast as its optotype [21]. The CSF Glare Tester, on the other hand, used sinusoidal gratings to measure contrast sensitivity with different contrast levels and spatial frequencies [49]. Furthermore, the type of glare sources differed, and the intensity of both glare sources were not disclosed. Hence, there is no basis to compare the two on illuminance.

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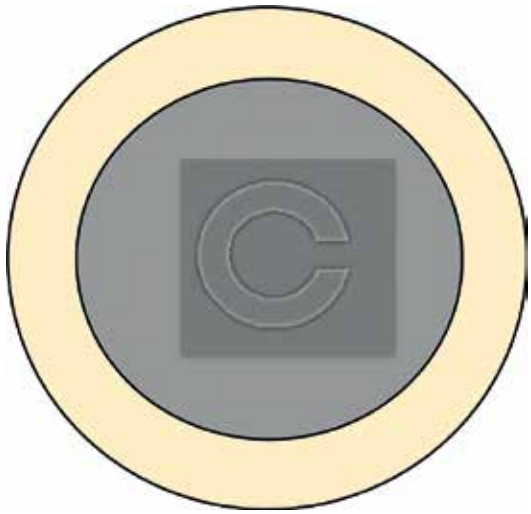

Figure 8. Schematic of the Ophthimus glare test with the ring light as the glare source and Landolt C at the center. Martin [49].

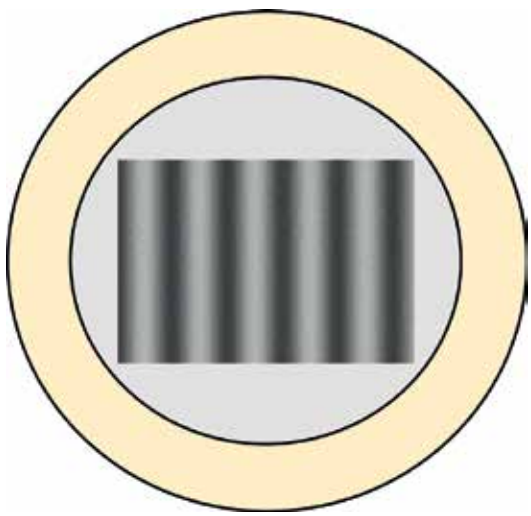

Figure 9. Schematic of Abrahamsson and Sjostrand glare device with a ring light as the glare source and sine wave contrast sensitivity in the center. Abrahamsson and Sjostrond [21].

When using the Landolt C, the protocol normally ensued a force choice answer. In the case of Ophthimus Glare Tester, the subjects were asked to report which direction the gap of the Landolt C was facing. This was done until the subject reached the lowest contrast in which the direction of the Landolt C could still be answered correctly [21]. This resembled the procedure of the CSF tester as the sinusoidal gratings were gradually increased to the contrast that was

barely visible to the subject under glare conditions. The task was done at all spatial frequencies [49]. Both glare test measured the lowest contrast level visible by the subject to determine their contrast sensitivity. These results were both used to calculate a glare score which was used to understand the visual function of the cataract subjects and ocular healthy subjects.

Their shared similarities in testing methods also yielded the same results where both glare tests displayed discriminative ability between cataract subjects and age-matched ocular healthy subjects. However, each study correlated their glare score with different measurements and so each drew their own specific inferences from their results. The Ophthimus Glare Tester study looked at cataract patients in preparation for cataract surgery. These individuals had normal visual acuity, but the results of the study showed their disability glare score to be significantly lower and they also reported visual complaints associated with glare. After the surgery, 24 out of 25 subjects had no self-reported glare problems but some of the subjects still displayed elevated glare sensitivity [21]. This supported the discriminative ability of the Ophthimus Glare Tester that the glare test could still distinguish between cataracts and ocular healthy individuals even after surgery when visual function improved. The validity of the Ophthimus Glare Tester's performance was supported by being relevant to the subjective visual complaints of the subjects as well as with the results of preoperative and postoperative surgery. The CSF glare tester, on the other hand, measured their scores against opacity levels of the cataract subjects. They demonstrated a correlation between the glare scores and the current pathology of each subject [49]. Hence, the validity that the CSF glare tester was based more so on physiological progress of the disease rather than subjective experiences. Both these glare tests exhibited strong discriminative findings but because the studies that utilized the tests based their results on different foundations, the information yielded by each glare testing device was distinctive. This also applied to the information each study provided about the effects of glare on cataracts even though the glare tests shared a number of similarities. And so more testing should be conducted to assess the comparative validity of these glare tests.

5. Conclusion

Functional vision deficits may occur in ocular healthy individuals and in individuals that have disease. It appears that glare testing can serve as a good indicator of visual function and may also be affected in disease states. As various new treatment modalities become available for age related macular degeneration, glaucoma and newer intraocular lens surgeries and laser refractive surgeries, treatment outcome may be better assessed using visual function tasks that are more difficult to perform and are more realistic of "real" world activities. To this accord a combination of glare testing with contrast discrimination may be well suited. The difficulties arise in lack of standardization of parameters or lack of existence of evaluation standards makes assessing of the glare tests very difficult. There is tremendous need for these standards setting and independent evaluation of these devices before a clinically acceptable standard can be obtained and accepted. It appears that although the glare testing shows huge promise it cannot be utilized clinically as a useful test and currently remains a technique useful for research arena.

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Section 3

Coping with Visual Impairment and Blindness

Chapter 5

Coping with Visual Impairment: Helping our Patients Face the Truth

Maynard McIntosh

Additional information is available at the end of the chapter

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Abstract

This chapter explores the factors influencing patients' ability to cope with imminent visual impairment (blindness) as well as methods that can be used to aid patients to rid stereotypes associated with their visual impairment. The factors that influence a patient's ability to cope with blindness can stem from both biological and social backgrounds; biological as it relates to age and social, meaning individuals that are in contact with the patient on a daily basis. Older patients tend to have more difficulty coping with blindness because they have established norms while children tend to feed of their parents' view of their visual impairment. Some patients practice a more spiritual approach to coping with their visual impairment. Although studies have been inconclusive; patients who pray and practice faith tend to have a more positive attitude towards their situation. Participating in sports can also help the visually impaired to have a more positive attitude towards themselves.

Keywords: visual impairment, blindness, coping, spiritual perspective, age

1. Introduction

Eye care professionals are faced with the formidable challenge and responsibility of being the voice of hope or doom for persons who desperately seek to cling to their last bit of vision. The history behind vision rehabilitation and integration of the visually challenged into society is an area of particular interest worthy of global collaboration [1]. The history of blindness, is a growing and now fashionable area of historical interest chronicled back to biblical era. The outlook has survived the mythological and demonic perspectives, thanks to science, and notable writers such as Helen Keller, John Milton and Louis Braille. While medical science

has unveiled the mysteries behind congenital conditions such as leber congenital amaurosis, glaucoma and its management, and the myriad of systemic diseases associated with degrees of avoidable vision loss; there is still great need to dispel myths about visual impairment.

Mills [1], attended a world conference on the history of blindness and listed some areas of discussion which summarizes historical perspectives and their evolution. The disciplines ranged from history, science, philosophy, medicine, social studies, religious history, to literature, art, and psychoanalysis. Papers included 'Different Ways of Seeing in the Middle Ages', 'Blindness, Learning and the Politics of Radio in Interwar France' and 'What a Blind Man Saw at the International Exhibition of 1862.

Visually impaired C. Kudlick, PhD (July 26, 2015) posited in an interview that most people think of disability as a biological, physiological thing that happens to someone, something ahistorical, rather than an identity shaped by history, politics, economics, society, and culture. She is now leading a charge to have the history of disability be a recognized field of study.

In line with the historical perspective of visual impairment, someone deemed as visually impaired often is condemned to a life of confinement and would not be deemed as 'valuable' to society, which often deters the coping process for patients due to lack of self-esteem. Therefore society plays a vital role in a patient's coping ability. Age also plays a vital role in the coping process, as older patients tend to have a harder time coping with their visual impairment. There are different methods for coping with blindness which will be discussed further in this chapter. Technology has over the years evolved and has helped doctors immensely in their prognoses and further help with 'breaking the news' of imminent blindness. Visually impaired patients can integrate into society and regain independence with help of a patient doctor and having accepted and adapted to their new way of life.

2. Age and its effect on adaptability

A paper-review study [2] found seven main areas impacted by vision impairment in adults over 60. These areas include: depression or mental health, anxiety, quality of life or well-being, social functioning, loneliness, or social support and interventions. It was concluded that most of these areas listed previously directly correlates to vision impairment (**Table 1**).

Older persons who have formed habits, and established norms will be more challenged by the impact on their lifestyle by impairment of vision function; some patients even remarking a preference for vision than any of the other senses. Many of my own patients remark they would rather deafness or even death itself, to blindness. Congenital and early onset vision impairment is less likely to lead to negative psychological outcome, and adaptability is more likely.

The attitude of parents towards their visually impaired children has a direct relationship to their child's acceptance of visual impairment. The attitudes range from shock and despondence, later to acceptance, overindulgence, overprotection, then comes the phase of hope in what medicine can do through the ophthalmologist. Parents have the opportunity and ability to mold the child in a positive attitude, and later acceptance by wider society.

Areas	Implications
Depression or mental health	Older people with vision loss are more at risk of reporting symptoms of depression and lower mental health, and being diagnosed with clinical depression than their sighted peers. Visual functioning rather than vision status has a stronger relationship with depressive symptoms.
Anxiety	Anxiety is not more prevalent and quality of life was not consistently reduced in persons with visual impairment.
Social functioning and Quality of life or well-being	Social functioning is likely to be reduced in individuals with vision loss, but not social network size or social activity.
Social support	Social support buffers against the presence of depressive symptoms but there is mixed evidence for social support to facilitate adaptation to vision loss and psychological well-being.
Loneliness	Loneliness is under-researched but one study suggested a higher prevalence of mild loneliness in individuals with vision loss.
Interventions	Interventions that address psychosocial needs directly are more effective than rehabilitation that addresses them indirectly through instrumental support.

Table 1. Conclusions drawn from paper-review study.

Parents initiate negative reactions based on their hopes and dreams of a normal child. Their social anxiety, which is fuelled by the embarrassment about the child's disability, is transferred to the child who likely feels differently. Simple activities like dressing, feeding, hygiene practices, play and study become a major problem from a parenting perspective. The fear and anxiety borne out of this parental attitude is what causes the child to experience the same in their interaction with their peers. Parents play an important role in stimulating a child's interest in his or her surroundings. Children become bored easily when the visual stimulus is withheld, hence parents must find alternative ways to drive the child's interest. "Remember that curiosity and desire arc the two eyes through which a person sees the world in its most enchanted colors" [3].

In planning the approach to counseling visually impaired persons, due diligence has to be given to those caring for them: their family members, loved ones, co-workers, and friends. Acceptance from family and the wider society is crucial to self -acceptance when considering physical disability. While parental attitude is critical in children, the attitude of adult support systems is important to adults facing visual impairment.

3. Breaking the bad news

Psychologists agree that the approach by a professional such as an Ophthalmologist in breaking the news of imminent visual impairment or eventual blindness is not unlike that for terminal illness or death; hence the phases of adjustment are similar [4]. The seven phases of adjustment to blindness are: (1) trauma (physical or social), (2) shock and denial, (3) mourning and withdrawal, (4) succumbing and depression, (5) reassessment and reaffirmation, (6)

coping and mobilization, and (7) self-acceptance and self-esteem [5]. The duration and outcome of the stages are unpredictable. The social and relational factors discussed in the previous section are among others that will influence the sequence.

Professionals must stress the positive in all situations, and outline to family members that all aspects of vision must be appreciated and respected. While 20/20 to 20/40 may be the gold standard to function vocationally in everyday life, many will come to appreciate the ability to distinguish light and dark, colors, large gratings and basic physical forms. Conducting acuity tests with the Berkley Rudimentary Vision Charts to establish and define Log Mar vision below the 20/400 line is a good start. At this point, patients with advanced glaucoma, diabetic retinopathy, age related macular degeneration, retinitis pigmentosa, and many others must realize the value of whatever residual vision they have. This will encourage compliance with the control of ocular and systemic diseases leading to blindness.

Patients deal with the sudden traumatic nature of the bad news with denial and disbelief. They view the abyss of isolation and torment socially [5]. Three factors that influence the severity and intensity of shock are: (1) the significance of the loss to the individual, (2) the suddenness or unexpectedness of the event, and (3) the degree of visual loss. Denial often lasts as long as the multiple opinions and therapies continue to change; when the eye care professional admits "we have done all we can do."

Tedrick [4] reports that self-pity is the main characteristic in the mourning and withdrawal phase. In this phase, individuals refrain from activities that they once loved to partake in. Tuttle and Tuttle [5] outlined the "D's" of depression as despair, discouragement, disinterest, distress, despondency and dissentment. The next stage in the adjustment process is reassessment and reaffirmation. In this phase/stage the client self-examines their life and finds meaning (purpose for living).

In the next stage (coping and mobilization), the patient will find information and resource that will support them in the change [4]. This phase has cultural overtones that determine how smoothly, and at what rate it progresses. In Jamaica, the go-to is the Society for the Blind and the philanthropic body that supports rehabilitation is the Lions Clubs spread across the island. As the patient crosses this phase into the final of self-acceptance, he/she gains self-confidence, and is able to accept the prognosis and move on with life. This may take a long time, and some may never get there.

4. Faith: influence of beliefs

In societies with strong theosophical backgrounds, the role of religious beliefs and claims to divine intervention are often brought to the fore. Patients facing imminent blindness look to prayer and divine healing in order to stay positive about their prognosis [6]. As a spiritual meditation, the activity has been shown to have physiologic benefits: clinically significant reduction in and ambulatory blood pressure, reduced heart rate, cardiorespiratory

synchronization promotion, altered levels of melatonin and serotonin, suppressed corticostriatal glutamatergic neurotransmission, immune response boosting, decreased levels of reactive oxygen species as measured by ultra-weak photon emission and reduced stress.

Invoking healing through prayer has been compared with the placebo effect, influenced by personality, expectancy, optimism, and motivation. Randomized controlled studies, have been inconclusive about the benefits of prayer, though faith has had documented positive impact on outcomes in treatment groups studied. Considerations given to the Hawthorne and Rosenthal effects have been attributed to some of these outcomes [6]. The Hawthorne effect refers to change that occurs as a result of the act of observation or measurement, whereas the Rosenthal effect refers to change resulting from observer or rater expectancy. Study conditions may foster exaggeration of measurement results on the part of the subject, and relaxation of the measurement criteria on the part of the observer respectively. With specific reference to glaucoma, I have found that patients with a positive attitude maintain functional vision despite advanced disc and Retinal Nerve Fiber Layer (RNFL).

Andrade and Radhakrishnan [6] examined the correlation of medical and psychosocial benefits to religious affiliations and practices by posing a series of questions (see Appendix 7.1). The results were inconclusive and as such a definite relationship could not be determined. The door to research of this nature remains open, with respect to religious and theological customs and beliefs.

Optimism in our patients facing blindness has a positive impact on the way they experience environmental stimuli, and how this information is used in coping strategies optimism may contribute to the well-being of persons who have lost their vision both directly and by promoting problem-focused coping and engagement in positive social comparisons [7]. Research has found that non-acceptance of vision loss, using avoidance or ventilation coping, and not facing reality with a positive outlook may lead to negative feelings and low adaptation. Ophthalmologists' attitudes towards the underlying disease, and how they convey hope or despair to patients, can influence psychosocial adaptation in patients. The concept of neuroplasticity and the individualistic nature of healing and repair cannot be minimized. Shifting the focus to realistic goals such as optimal IOP in glaucoma, and discussing breakthroughs in neuro-therapy such as stem cell research will help keep hope alive and foster positive outlook.

5. The second opinion

Murphy [8] writes of her experience with a poor visual prognosis, where second opinion changed her life in a day, from despair to hope. An ophthalmologist diagnosed her sudden onset of loss of vision as macular hemorrhage of questionable etiology, giving a 2-week period before anticipated total vision loss. Her second opinion from a younger eye surgeon (and has a good prognosis), gave a diagnosis of Central Serous Maculopathy which is self-limiting. Patients are entitled to this review, and doctors should have no insecurities about this, as medical research is very dynamic.

Diagnosis of irreversible (legal) blindness should be made by medical or eye care professionals qualified to make such pronouncements, and the same should be validated by objective methodology as far as is possible. There are legal ramifications to unfavorable visual diagnoses with socio-economic, emotional, vocational, relational, and psychological consequences. Pronouncements should be made in the presence of a legal support system, as well as the full cadre of relevant eye care professionals.

Case in point: Macular holes are common, and when greater than 600 microns, have a poorer prognosis for repair and vision rehabilitation. This, until recently with the advent of autologous blood as an adjunct to macular hole surgery. Research continues to change the prognoses for vision rehabilitation in many cases traditionally considered end-stage. Informed second opinion should be a part of due process in the management of patients with unfavorable visual prognoses that can negatively affect their quality of life [9, 10].

The prognosis of conditions like glaucoma, retinitis pigmentosa and macular degeneration, to name a few are largely subjective. Advice on prognosis and related patient counseling needs to be collaborative, involving psychologist, ophthalmologist, patient, care-givers, and key opinion leaders who can use latest investigative and therapeutic tools to alter the course of progression where possible. Acceptance of a diagnosis and prognosis had a large cultural and socio-economic component that differs significantly according to the cultural milieu [11]. Second opinion in many cases may involve the use of tele-medicine, research, and professional exchanges across borders.

6. Role of sports as a coping mechanism

It is a well-established fact that physical activities including sports, build self-esteem and self-acceptance, as well as enhance physical, mental, and emotional health [12]. This has been known to medicine as far back as B.C. 460 (alluded to by Hippocrates). The International Blind Sports Association (IBSA) is the arm of the Paralympic Movement governing professional and competitive sports for the visually impaired. British athletics in schools was used as an inspiration for developing Paralympic sports, Para (parallel) – lympics (running just after the regular Olympic program) [11]. The running, jumping, and throwing activities were found to boost not only physical wellbeing, but brought inclusion and equality to persons otherwise marginalized due to disabilities. Quoting from the Paralympics manual:

> *'The fundamental principle guiding the Paralympics movement is that elite level athletes with physical disabilities should have opportunities and experiences equivalent to those afforded elite athletes without disabilities…Competitive sports have proven to be an effective vehicle to promote equality, inclusion, accessibility and awareness about the capabilities of those with physical disability…and dispel the stigma surrounding disability and illuminate the realm of possibility…"* [13].

Wanderi [12] looked at the role of sports in the lives of Kenyans in educational institutions and communities on a whole. The Government of Kenya in 1988 outlined a decade plan for the integration of education to the tertiary and post graduate level, where both physically challenged and those desirous of working with the disabled would receive equal training in the universities, a program which spread across academic and sports departments. The 2006–2011

strategic plan of the Ministry of Education in Kenya was a highlight of Kenya Government's commitment to improving the fate of the disabled. Legislations and Governmental policies need to stand behind the inclusion and empowerment of visual impaired in their societies, thus minimizing the sense of dislocation and hopelessness of those with visual, and other physical impairment.

The ability of sports to unite communities, regions, and cultures has been demonstrated time and again, and is even more powerful when viewed from the standpoint of disabilities. There is an added measure of respect and dignity that transcends the winner's podium. Just participation in itself lends to self-confidence, respect from community, and aids the rehabilitative efforts of those caring for the visually impaired. Physical activity among the visually impaired enhances coordination as well as sharpens proprioceptive and other senses that are needed in daily life. There is a bonus in other areas of health that would be threatened by a sedentary lifestyle that can result from the loss of self-confidence and independence accompanying significant vision loss.

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Chapter 6

Psychosocial Adaptation to Visual Impairment

Yukihiko Ueda

Additional information is available at the end of the chapter

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Abstract

Acquired visual impairment evoked several psychological reactions. A person's adaptation to these reactions and their associated characteristics such as degree of vision loss, membership of a specific demographic, and impact on personality had been investigated. Socio-environmental variables also had an impact on psychological adjustment, and adaptation to vision loss had been explained using psychoanalytic models and stage models. Previous research had thus developed adjustment models incorporating the variables of anxiety, depression, self-esteem, self-efficacy, locus of control, acceptance of disability, attitudes toward blindness, and attributional style, among other influences. However, effective types of treatment based on these variables had not been empirically demonstrated. While grief work had been commonly used in intervention strategies, and there was now more information available about group counseling in this field, their evaluation had been insufficient. As an alternative, we implemented a structured group counseling program to decrease psychological distress in adults with visual impairment. The results indicated that participants who engaged in individual therapy in addition to group counseling showed decreased depression, fatigue, and confusion, and a significantly improved acceptance of their disability. The group counseling combined with individual cognitive therapy could be the effective tool to improve social influences and internal self of the person with visual impairment.

Keywords: visual impairment, rehabilitation, adaptation, group counseling, cognitive therapy

1. Introduction

It was known that acquired visual impairment evoked several psychological reactions, including shock, depression, grief, resentment, shame, self-derogation, a resigned attitude, feelings of inadequacy, and feelings of excessive guilt (e.g., see [1, 2]). People's reactions to visual impairment were lack of understanding, misconceptions, and/or prejudice. In this chapter, we

focused on the acquired visual impairment that occurs in adults. In this discussion, we provided an overview of the theories and empirical studies relating to the psychosocial problems that were commonly experienced by the visually impaired.

1.1. Causes of visual impairment

Visual impairment might result in serious difficulties, because human beings depended on visual perception to get most of their information from the world around them. It might also trigger a psychological crisis that could promote an intention to seek "death," as Carol described [3]. In Japan, an approximately 310,000 people suffered from visual impairment. However, this number was only those who had a certified disability; there were more people suffering from visual impairment than were on official lists. Visual impairment was brought about by various causes such as eye disease, systemic disease, encephalopathy, and traumatic injury. Eye diseases include glaucoma, retinitis pigmentosa, optic atrophy, macular degeneration, retinopathy of prematurity, and so on, while systemic diseases include diabetic retinopathy and Behcet disease. Encephalopathy includes visual impairment caused by brain injuries, and postoperative impairment from brain tumors.

The aspects of psychological distress resulting from these situations were different, depending on the time of onset and the type of visual impairment experienced.

2. Psychosocial problems

Though visual impairment might differ depending on country and age, there was still a paucity of empirical research concerning psychosocial adaptation to acquired visual impairment [4, 5].

2.1. Demographic variables, degree of visual loss, personality, and adaptation

Some studies had investigated client adaptation to these problems, as well as the associated features of clients, including their demographic and personality characteristics, and their degree of vision loss. Though some studies showed that psychological reactions differ depending on the degree of visual loss, the studies, which suggest that there was a positive correlation between residual vision and adaptation, were those by Fitzgerald et al., Lukoff and Whiteman, and Wulsin et al. [6–8]. Against these studies, Teitelbaum et al. failed to find such a relationship [9].

Bauman examined the relationships between psychological adaptation and a client's visual, medical, personal, social, educational, and vocational histories, through structured interviews incorporating a comprehensive test battery [10]. The segmented data from this study showed that their well-adjusted group (37% of 400 persons) was (a) independence, (b) mostly mobile, (c) maintained satisfactory home and community activities, and (d) had a successful work history. On the other hand, their identified maladjusted group (29%) was (a) dependence, (b) mobility-dependent on others, (c) engaged in only limited home and community

activities, and (d) had no recorded work history. They were also able to show that the well-adjusted group demonstrated higher scores on intelligence, manual dexterity, emotional stability, and realistic acceptance of their visual impairment, and attained higher educational levels than the maladjusted group. However, no differences were found between these two groups on the degree of vision loss, health indices, or the level of social interaction. A follow-up study carried out 14 years later showed that these characteristics had been retained [11].

Joffe and Bast examined the relation of ego functioning and adaptation of 101 men with a visual impairment using the California Psychological Inventory (434 items' questionnaire that include 18 scales. Each scale measures interpersonal adequacy, character, intellectual efficiency, interests, etc.) and extensive structured interviews [12]. In this study, occupational status and mobility were used as the index of adaptation. No differences were found between the employed and the unemployed groups on measures such as educational level, age, degree of vision, and several psychological attributes such as defense and coping. However, by combining occupational status with mobility, the study examined the differences between accommodators (employed and high-mobility skills) and non-accommodators (unemployed and poor mobility skills). The researchers found that accommodators used extensive coping strategies, such as mature, adaptive, flexible, purposive, present-oriented, and reality-based behaviors. Accommodators also included objectivity, intellectualization, suppression, and tolerance of ambiguity as techniques to overcome visual impairment problems. By contrast, non-accommodators tended to rely on defensive strategies such as immature, non-adaptive, rigid, past-oriented, and irrational reactions, and used projection, regression, fantasy, displacement, rationalization, and doubt in their reactions.

2.2. Individual factors of emotional and psychological reactions to visual impairment

The individual variables that acted in specific situations to exacerbate or reduce the differences in each of these reactions, and determined the degree of further psychological adaptation, had been explained from the various viewpoints of different schools of thought. A sketch of this material is discussed in the next section.

2.2.1. Psychoanalytical and psychodynamic models

One of the earliest theoretical approaches that emphasized the importance of vision in personality development and later adult life was developed by psychoanalysts. Blank maintained that reactions to visual impairment could often be traced to the unconscious significance of (1) the eye as a sexual organ, (2) the eye as a hostile, destructive organ, and (3) blindness as a punishment for sin (like castration) [13].

Traditional psychodynamic models emphasize the importance of concepts such as loss, mourning, and grief. In this context, Caroll maintained that the losses forced on the blinded person were many [3]. They interlocked; they overlapped one another. Any one of them was severe enough in itself. Together, they made up the multiple handicaps that were blindness. Each loss involved a painful farewell (a "death"), then, and Caroll identified 20 types of losses from blindness in his classic writing. These were (loss of) (1) physical

integrity, (2) confidence in the remaining senses, (3) reality contact with environment, (4) visual background, (5) light security, (6) mobility, (7) techniques of daily living, (8) ease of written communication, (9) ease of spoken communication, (10) informational progress, (11) visual perception of the pleasurable, (12) visual perception of the beautiful, (13) recreation, (14) career, vocational goal, job opportunity, (15) financial security, (16) personal independence, (17) social adequacy, (18) obscurity, (19) self-esteem, and (20) loss of total personality organization. Since a blind person lost such a lot, Caroll emphasized that rehabilitation had to provide effective substitutes.

2.2.2. Stage model

Caroll's loss model had been further developed. A stage model, which insisted that emotional psychological reactions to trauma occur in stages, was one such development. This theory explained that the psychological reactions experienced after acquired visual impairment might pass through the stages of (1) shock, (2) depression, and (3) recovery [1, 13]. Another stage model proposed by Allen suggested three adaptation processes: (1) pre-impact phase, (2) impact phase, and (3) learning to live with impairment [14].

Related to the stage models, there were some studies of the impact of denial on the process of adaptation. Dover recognized the importance of denial as a defense to ward off anxiety [15]. She emphasized that denial was frequently manifested through a search for new medical discoveries and magical treatments. Shulz, in distinguishing between denial of the severity of the condition and denial of the affective content or meaning of the visual loss, considered that the latter could interfere with the process of adaptation [16].

3. Personality and adaptation

3.1. Anxiety

There were some studies that have focused on the relationship between personality traits and adaptation to visual impairment. These studies were called disposition (or trait) theories. The first personality trait that affected rehabilitation was anxiety. As stated above, Dover recognized that denial was a defense to ward off anxiety; in the phase of anxiety, people with visual impairment often did not participate in rehabilitation, or reject to participate. They denied visual impairment of themselves because of anxiety [15]. Moreover, anxiety resulted in lowered attention spans and decreased the ability to use cues of environment, influenced learning, and performance in personal rehabilitation [17]. In addition, the learning of the person with high anxiety was slower, and the retention of what was learned was less [18]. That was why treating anxiety was significant in rehabilitation for visual impairment in which Braille, mobility techniques, and the techniques of daily living were acquired.

3.2. Self-concept and self-esteem

Self-concept was defined as "a collection of thoughts and feelings one about oneself," [19] and self-esteem had been used as an affective dimension of the self-concept [20]. The reason that

self-concept was related to rehabilitation and adaptation was that human beings tend to initiate behavior to reduce the discrepancy between "the present self" and "the ideal possible self" [21], and seek feedback that was consistent with their self-concept, but avoided information that was contradictory [22]. Therefore, the self-concept idea related to whether the individual with visual impairment could initiate new behavior and how he/she could adjust to their new life. In other words, psychological reactions might differ depending on how much his/her self-concept was defeated. If a person shaped negative self-concept by acquiring visual impairment, and resisted change, it could become very difficult to advance toward the goals of a more independent self in rehabilitation [23].

Tuttle had produced many examples of possible discrepancies that persons with visual impairment might encounter between the way they saw themselves and how they were seen by significant others [20]. These discrepancies needed much effort to resolve and had major impacts on their personal adjustment [23]. This was why there were some studies that had made considerable efforts to establish empirical evidence of the differences in self-concept and self-esteem between people with vision loss and sighted people [24]. However, these studies produced contradictory results because they involved confounding variables, such as inappropriate measuring instruments, the length of time that people with visual impairment had experienced vision loss, and the diverse range of coping strategies they used [23]. Still, self-concept and self-esteem were not closed traits in an individual. Also, their relationships with the people around them should not be ignored. We will discuss this issue later.

3.3. Self-efficacy

Another concept that is related to rehabilitation and adaptation was self-efficacy. In the past, the concept of confidence and motivation was widely used in the field of rehabilitation, because one of the problems rehabilitation personnel had been struggling with was low confidence or unmotivated clients. However, since the widely used concept of motivation was so simple, Dodds outlined the usefulness of the concept of self-efficacy for rehabilitation [25]. The concept of self-efficacy, originally proposed by Bandura, was based on the notion that "our expectations have effect on our motives and behavior" [26].

Moreover, Dodds et al. developed an adjustment structural model comprising anxiety/depression, self-esteem, self-efficacy, locus of control, acceptance of disability, attitudes toward blindness, and attributional style by using the LISREL (linear structural relation) model (the statistical methods to formulate theoretical model for manifest variables and latent variables from collected data) [27]. Additionally, two factors, which were not assessed directly but appeared in latent form, were identified. These factors were "self as agent" and "internal self-worth," and seemed to explain the high interrelationships found among above seven factors assessed by the adjustment structural scale (**Figure 1**). Based on this, Dodds et al. asserted that successful adaptation was multidimensional and includes (1) low levels of anxiety and depression, (2) high levels of self-esteem and self-efficacy, (3) a high sense of personal responsibility for recovery, (4) a positive attitude toward visual impairment, and (5) acceptance of one's own visual disability. They also suggested that the process of adaptation was inclusive of changes in both negative aspects (decreasing anxiety and depression) and positive aspects (improvement of self-concept, sense of mastery, and self-control) [27].

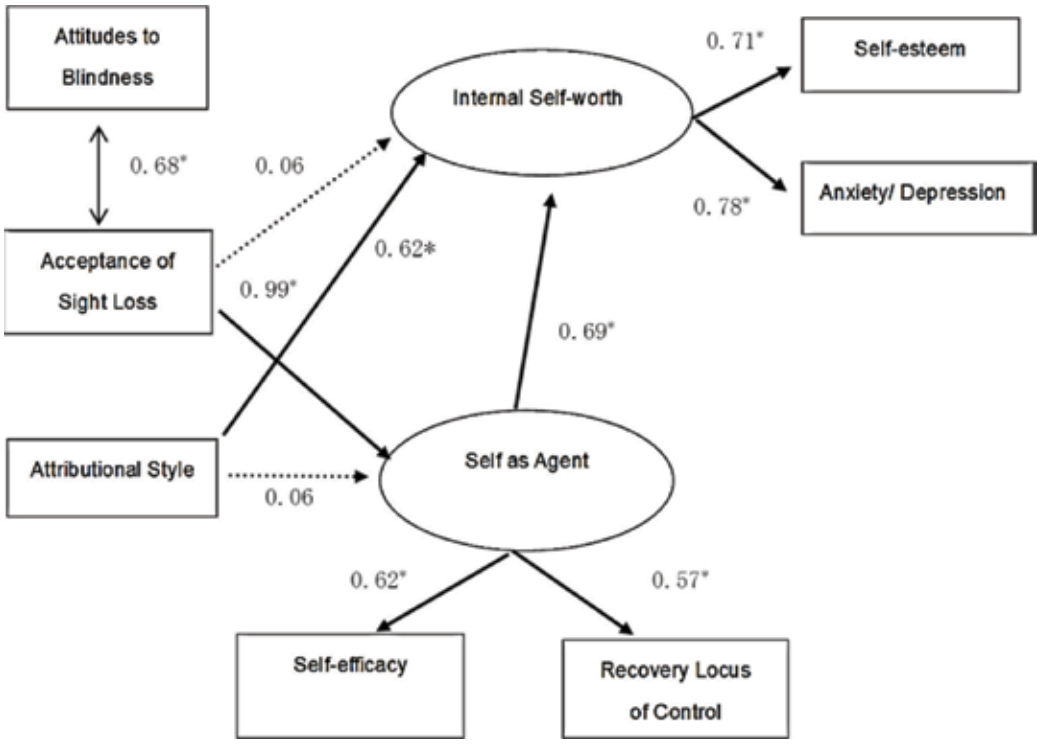

Figure 1. LISREL structural model with path coefficients (*$P < 0.05$). Reprinted with permission of the publisher from Dodds et al. [27]. Copyright © 1994 by American Foundation for the Blind. All rights reserved.

Furthermore, based on the results of the LISREL analysis, they suggested that these factors might influence rehabilitation practice. For example, attritional style had a direct effect on "internal self-worth," and "internal self-worth" was related to anxiety/depression and self-esteem. Acceptance of sight loss was also strongly related to "self as agent." "Self as agent" reflects a belief in one's ability to control future goals and tasks, was related to self-efficacy and recovery locus of control, and directly related to "internal self-worth." From these results, Dodds et al. concluded that, while counseling might not directly lead to measurable improvements in self-worth, it might improve the motivation for an individual to act in ways that brought about successful adjustment outcomes [27].

4. Social factors

4.1. Socio-environmental influences

These theories and studies mentioned earlier had been developed from the standpoint that emotional and psychological reactions and adaptation varied according to the individual factors. However, there were studies from the alternative standpoint that the reactions of individuals with visual impairment were the consequence of influence from other people. Thus,

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socio-environmental influences such as prejudice and interactions with others also had an impact on psychological adjustment.

Cutsforth, a blind psychiatrist, was the first to assert the influence of society on such reactions [28]. He stated that the characteristic of emotional disturbances was that they were evoked from the social situations that blindness created and not from the sensory deprivation itself. Thus, it was difficult to find any evidence that blindness itself was productive of emotional disturbances. Chevigny and Braverman described that society included the beliefs, feelings, and consequent expectations that sighted persons inflicted upon the blind person [29]. They stated that every blind person reacted to this environment, either consciously or unconsciously, and resentment was a primary reaction that emerged from this social situation. Thus, a major issue in other people's reactions to blindness was "lack of understanding and misconceptions." The core of the self-concept of persons with visual impairment was the stereotypical attitudes and expectations of sighted people around them, which became the rules by which persons with visual impairment sculpted their thoughts, feelings, and daily actions. It was thus impossible for persons with visual impairment to ignore these beliefs, and they had no choice but to respond to them. The result was a feeling of shame, inadequacy, and self-derogation [30, 31].

Though empirical studies had been conducted to discover how other people regard visual impairment, the findings were confounded. This was caused by the variety of using psychological tests, different eliciting stimuli, and confusion regarding whether the research was measuring people's attitudes toward visual impairment, or toward people with visual impairment [32]. However, many of the confusing and discrepant findings suggested that people's attitudes toward visual impairment were not unidimensional, but were instead multidimensional [23]. Whiteman and Lukoff [33] identified five dimensions of other people's attitudes toward visual impairment or people with visual impairment: (1) personal attributes reflecting a negative view of the emotional life and general competence of people with visual impairment; (2) social attributes reflecting a readiness for interaction with people with visual impairment, and a positive view of the social competence of people with visual impairment; (3) evaluation of visual impairment that showed the degree to which visual impairment was perceived as threatening and uniquely frustrating to one's self and others; (4) non-protectiveness, reflecting a lack of protectiveness and sympathy; and (5) interpersonal acceptance, reflecting an emotional acceptance of people with visual impairment in interpersonal situations.

In addition to these studies, there were researchers who insisted that the attitudes of other family members had a strong impact. Versluys [34] and Featherstone [35] noted that families who communicated positive attitudes helped stabilize their visually impaired relative's self-concept. Large noted that the attitudes of family members, especially parents, had a powerful influence and, depending on whether the influence was positive or negative, had lasting effects [36]. The reason why the family had such a strong influence was that it served as the major source of interpersonal influences that affected what visual impairment meant to the person suffering from this, and what he/she did with it [37].

Vision loss was likely to cause disturbances in an individual's balance between independence and dependence. The affected person had to now depend to a greater degree on others to

assist him/her in performing the many tasks of daily living and especially for help in travel until he/she learned techniques for functioning without vision. Therefore, this could lead to serious problems if an individual had dependency conflicts throughout his/her life [23]. Thus, we needed to understand the influence of the family from the viewpoints of dependence and independence, and the roles in the family formerly occupied by the visually impaired person that had changed.

4.2. Combination of individual and socio-environmental factors

There were some researchers who attached great importance to both individual and socio-environmental factors in addition to these socio-environmental variables. Bauman and Yoder, for example, recognized the impact of both the situation and the reaction of family members, friends, and medical personnel on the adjustment of newly visually impaired person [11]. Roberts stated that visual impairment presented the human organism with one of the most sweeping environmental adaptations conceivable, and the views of other people made it necessary for the visually impaired person to reexamine and often to redefine his/her own self-concept as well as his/her previously established roles and procedures [38]. Yeadon and Gryson stated that reactions to visual impairment steamed for two main sources: the attitudes of the person and others, and the age at the onset of visual impairment [39]. Tuttle also thought that both "physiological loss of vision" and "society's prevailing attitude" had an equally important impact on the reactions of an individual [20].

4.3. Prejudice by interaction with others

Many visually impaired people were stigmatized when they were living in various locations. In this context, the stigma is referred to "some deviation from a norm or standard" [23]. Persons with visual impairment were often stigmatized by their appearance or by the equipment (white cane, special glasses) they used. The problem of stigma, however, did not reside in the person who possesses the stigma, nor in the persons who reacted to the stigma, but in the interaction between them [40, 41]. People who were stigmatized elicited atypical reactions and behaviors from the public. These reactions could impact on the self-concept of the person with the disability, as well as on his/her behavior [23].

Understanding these interactions, Barker et al. suggested the concept "A new psychological situation" [42]. This was a concept that covered the fact that a person would engage in behaviors that attracted and repelled, trial and error, and experienced frustration, then withdrew to the safety of the old in the situation where the location of positive goals and the path by which they could be reached were clear. Visually impaired persons frequently experienced this "new psychological situation" when lacking a necessary tool for dealing with the situation, or when confronting the reactions of others.

Another concept that might be important was the one of "overlapping roles." If the person with visual impairment had no problems in their limbs, they might find themselves more torn between the roles of independent and dependent than the persons with physical disability. This condition might result in the feeling of being a "marginal man," and this "marginal individual"

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might have ambivalent feelings about his/her new identity, such as "Am I an independent person or a dependent person?," and might wish to reject it.

Then, there was the concept of "passing," which often affected persons with low vision, not those with total visual impairment. They were passed up the fact of vision loss by people around them in almost all living situations. This might have an impact on the identity of the person with low vision and on the decision of when to disclose their visual disability.

5. Interventions for psychosocial adjustment

As described earlier, the difficulties in, and the complexity of, the psychological adaptation of people with visual impairment were caused by the interaction of personal factors, environmental factors, and individual behavior. Therefore, the methods of support intervention had also varied, depending on the differences in the understanding of the cause of psychological reactions or of the difference of the purpose of the intervention.

5.1. Individual psychotherapy

5.1.1. Grief therapy

Treatments that were effective against the psychological reactions to acquired visual impairment had not been widely empirically demonstrated [4]. Grief work had been one commonly used intervention strategy, although empirical evidence of its efficacy is lacking.

Choldon stated that the therapist should be a relatively fixed, nonthreatening, and warm figure [1]. It was fruitful to point out to a patient conscious side which he/she did not accept the disability, and situations where they could avoid the difficulties that visual impairment brought. But as, in the shock stage, any readjustment effort was not effective, it would seem unwise to do so. After this stage, the reactive depression stage began, which was a period of mourning for their eyes, in which the patient had to die as a sighted person and was an important and necessary phase in the reorganization process. The patient needed to experience this depression before they could accept the reality of visual impairment; efforts to prevent or abort it should not be made. In this stage, it was better to let them alone to cry. After this, it was possible to hasten the movement out of the depressed state by the judicious use of activities and training tasks in the rehabilitation. Having successfully accomplished some task believed difficult at first might lift their mood, but if an overambitious task that they could not accomplish was presented, their depression might be intensified.

5.1.2. Cognitive therapy: maladjusted belief

Needham and Ehmer categorized the 16 maladjusted belief statements that visual impaired people often made into four categories [43]. These were that (1) blind people were different from sighted people in their self-worth and value (e.g., an individual's worth was dependent upon his/her physical adequacy. So, blind person was of little value); (2) blind people had a

unique psychological constitution (e.g., blind people had to be either gifted or defective in their intellectual functioning); (3) blind people had a special relationship with other people and society in general; and (4) there were magical circumstances about blindness (e.g., blindness would be cured by a new scientific discovery, or new products of engineering would solve the problems of blindness).

Additionally, they suggested that maladjusted beliefs mentioned earlier about blindness could affect and limit the lives of visually impaired people. The mythologies and irrational beliefs about visual impairment were part of our general culture and were just as prevalent among sighted as among visually impaired people. This meant, therefore, that any single irrational belief statement that had a potential to cause much unhappiness for a visually impaired person would become the self-defeating thought that could limit their living. Hence, the appropriate intervention included uncovering an individual's irrational beliefs by direct inquiry, knowing their peer group's different capabilities, and seeking views on their impairment in rehabilitation settings or in psychotherapy.

Within the therapeutic milieu (rehabilitative or psychological), the person with visual impairment should be regarded as a student and a learner rather than as a patient, be contacted with objective and realistic attitudes of their condition, and be able to test the accuracy or inaccuracy of their own beliefs. It was possible to bring about cognitive changes through such interventions. Modifying maladjusted beliefs by effective intervention (cognitive therapy) and acquiring coping techniques for visual impairment (e.g., mobility technique, Braille, daily living technique) would make it possible for visually impaired persons to engage in a limited but many activities, to have dreams that lend themselves to real fulfillment, and to experience the pleasures that were indeed possible even though they were blind. It was important for the therapist to know what mythology existed and what could be accomplished to change this through rehabilitation.

5.1.3. Group counseling

In contrast to the paucity of information regarding the efficacy of individual psychotherapies for the visually impaired, more information was available regarding the usefulness of group counseling. In general, group treatment approaches had been more effective than individual treatment [44]. However, group treatment had been even more effective when used concurrently with individual counseling [45].

This tradition began with Choldon [1], followed by Herman [46], Ross and Anderson [47], Goldman [48], Manaster [49], Roessler [50], and McCulloh et al. [51]. Group counseling approaches were not uniform, however, and had different theoretical bases, goals, and intervention techniques. Moreover, there could be differences in economic status and educational level between clients. These differences could influence original adaptation level of the clients. Furthermore, in nearly all the studies, outcomes were evaluated based solely on therapists' impressions. Only Roessler used a control group while evaluating outcomes using objective indicators such as the standardized self-esteem scale and the locus of control scale [50]. The structured group counseling that he named "Personal Achievement Skills Training (PAS)" included communication skills, value

clarification, problem-solving skills, and self-modification. Control group participants were involved in group counseling focusing on personal feelings and adjustment to disability. PAS group ($n = 16$) and control group ($n = 18$) were randomly assigned. The participants in PAS group showed significantly higher self-esteem and goal achievement than the control group.

Van der Aa et al. conducted a meta-analysis of 22 studies of psychological intervention implemented between 1981 and 2015 [5]. These studies included group-based cognitive-behavioral intervention, self-management programs, problem-solving treatment, psycho-education, relaxation training, and behavioral activation. Fourteen studies were randomized control trials (RCTs), while in 15 studies, the participants' mean age was over 60. The studies demonstrated that intervention reduced depressive symptoms significantly, but anxiety symptoms, mental fatigue, psychological stress, and psychological well-being were not improved significantly. Given the higher age of participants, the effects on depressive symptoms, psychological stress, and psychological well-being were small.

5.1.4. Structured group counseling combined with individual cognitive therapy

Considering the interaction of personal factors, environmental factors, and behavior in the psychological adaptation of persons with acquired visual impairment, we implemented a structured group counseling program as part of a Living Skills Training Program [52]. A quasi-experimental study design was used to compare the outcomes of a 6-month group counseling program with and without individual cognitive therapy, which included a control group. The group counseling program was based on weekly 90-min sessions consisting of three components: (1) talking about experiences and feelings; (2) psycho-education about disability, eye diseases, and social resources; and (3) relaxation and meditation.

Individual cognitive therapy was scheduled once a week for 45 min. Clients talked mostly about how they felt and what they thought during the group counseling sessions, and compared other group participants' responses to their own expectations and beliefs. When distorted negative cognitions were identified, these were explored and modified during individual therapy sessions based on the actual attitudes of the other participants.

The control group was taken from the participants of the Living Skills Training Program. This program consisted of orientation and mobility training, writing and reading Braille, house cleaning, cooking, sports, recreation, and computer training. The programs were held 5 days per week, and the course ran for 6 months. The results indicated that participants in skills training alone ($n = 32$) improved significantly in acceptance and attribution style, while also showing a trend of improvement in tension anxiety and self-esteem. However, participants with high levels of psychological distress (who indicated a *T*-score of Profile of Mood States Test over 60, $n = 10$) did not show any such improvements. Nevertheless, highly distressed participants that participated in group counseling ($n = 18$) showed significant improvement in their attitudes toward visual impairment and reported decreased anxiety. Moreover, participants that chose to engage in individual therapy in addition to group counseling ($n = 9$) also showed a decrease in depressive mood, fatigue, and confusion, as well as significantly increased acceptance of their visual disability.

5.1.5. Case example

Mr. O was a 34-year-old man who had acquired a visual impairment due to pigmentary retinal degeneration and had participated in the Living Skill Training program. Before participating in this program, he had attempted suicide. At first, he had a thought that "there is no meaning in the life with loss of vision." As treatment progressed, he began to clearly recognize the negative beliefs regarding his disability such as "people think that visually impaired person can do nothing." Hearing his peers' thoughts in group counseling, experiencing others attitudes in mobility training, and through cognitive modification in individual therapy, he could modify negative beliefs and have positive beliefs, such as "I'm not so poor." and "I don't want to be back to healthy person. I'm OK as I am now." The outcome was that there was improvement in his depression, attitudes, and acceptance of his disability. After this program, he began to live independently as a practitioner of acupuncture [53].

6. Conclusion

Psychological distress and reactions to visual impairment were caused by the interaction of personal factors and social factors. Therefore, it was difficult to solve these problems by single intervention. Although acquiring independent skills was effective for psychosocial adaptation, group counseling combined with individual cognitive therapy could be the effective tool to improve social influences and internal self of the person with visual impairment.

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Chapter 7

Assistive Systems for the Visually Impaired Based on Image Processing

Hotaka Takizawa and Mayumi Aoyagi

Additional information is available at the end of the chapter

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Abstract

In this chapter, we proposed three assistive systems for visually impaired individuals based on image processing: Kinect cane system, Kinect goggle system, and light checking system. The Kinect cane system can detect obstacles of various sizes and also recognize objects such as seats. A visually impaired user is notified of the results of detection and recognition by means of vibration feedback. The Kinect goggle system is another type of wearable system, and can make user's hands free. The light checking system is implemented as an application for a smartphone, and can tell a visually impaired user the ON/OFF states of room lights and elevator button lights. The experimental results demonstrate that the proposed systems are effective in helping visually impaired individuals in everyday environments.

Keywords: assistive system, Kinect, cane, goggle, smartphone, camera, image processing, obstacle detection, object recognition, light checking

1. Introduction

The world health organization estimated the number of visually impaired individuals to be approximately 285 million in 2014 [1]. Many of them use white canes to detect obstacles around them. However, the detectable ranges of white canes are very short. Guide dogs are also used to navigate visually impaired individuals to their destinations while avoiding obstacles. However, it is difficult to provide the sufficient number of guide dogs due to long-time periods and expenses to train them. In order to overcome these problems, extensive research has been dedicated to creating assistive systems for the visually impaired [2].

Obstacle detection is one of the representative research themes. Many research groups have proposed obstacle detection systems based on laser sensors [3–11], single charge-coupled

devices cameras [12–16], ultrasonic sensors [17–29], stereoscopic cameras [30–41], or RGB-D cameras [42–46]. These assistive systems are built on the basis of the concept of the electronic travel aid (ETA) [47, 48], which aims to assist visually impaired users in walking while avoiding obstacles. Therefore, these systems can notify the users about obstacles but cannot tell them what kind of objects they are.

Here, let us consider a situation where there is a seat (bench) in front of a visually impaired user as shown in **Figure 1**, and the user wants to sit on the seat to take a rest. In this situation, the seat is not just any obstacle, but a useful equipment. If the user uses one of the obstacle detection systems mentioned earlier (see **Figure 1(a)**), he or she is required to confirm the obstacle by himself or herself. However, if the user uses an object recognition system, which can determine the object to be a seat (**Figure 1(b)**), the user can obtain a benefit. It is necessary to build an assistive system to recognize objects around a visually impaired user.

Several research groups have proposed object recognition systems. Drug packages [49], classroom doors [50–53], podiums [50], and pathways [54, 55] are recognized by using barcodes [49, 56], augmented reality markers [50, 52–54], radio frequency identification tags [23, 51, 57, 58], Bluetooth devices [59], wireless network devices [55], or visible light communication devices [60, 61]. These physical devices are useful, but it is difficult to deploy such devices in everyday environment.

Other research groups have also proposed assistive systems to notify visually impaired users about tables [62], color blocks [63], and staircases [64–67] by means of laser range sensors [65–67] and Kinect sensors [62, 63]. These systems are useful, but are not sufficient yet. Other types of object should be recognized to help visually impaired individuals more.

This chapter proposes our assistive systems not only to detect obstacles of various sizes but also to recognize objects of various types by use of image processing technique.

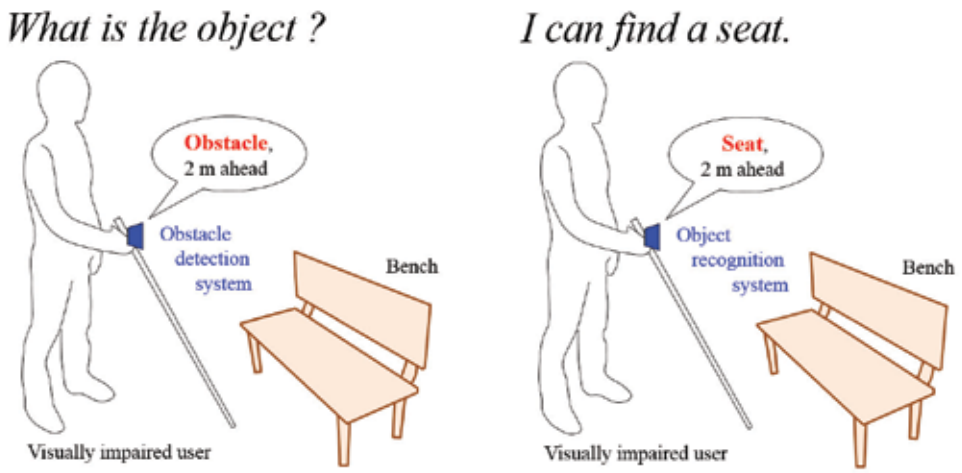

Figure 1. Obstacle detection system (a) versus object recognition system for the visually impaired (b).

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2. Kinect cane system

Figure 2 shows our Kinect cane system composed of a white cane and a backpack [68]. A Kinect sensor, a numeric keypad, and a tactile device are attached to the white cane. Kinect is an infrared-based range sensor for a consumer game machine, that is, Microsoft Xbox, and the white cane is also a commercial product for the visually impaired. (The sensor and the cane are approximately 300 and 100 USD, respectively.) The Kinect sensor is set at 75 cm from the floor. These devices are connected by wires with a portable personal computer and a UPS battery in the backpack. The computer and the UPS battery are used for device controlling and power supply, respectively.

In this system, the X, Y, and Z axes of the world coordinate system are defined to be the horizontal vector oriented from left to right, the vertical vector oriented from top to bottom, and the horizontal vector extending from a Kinect sensor into the environment, respectively.

The Kinect cane system can detect obstacles and recognize several objects, such as seats, by use of our special computer programs implemented for the following methods.

2.1. Obstacle detection

The Kinect cane system can detect obstacles which would prevent a visually impaired user to walk safely [45]. We provide two detection methods of small and large obstacles considering

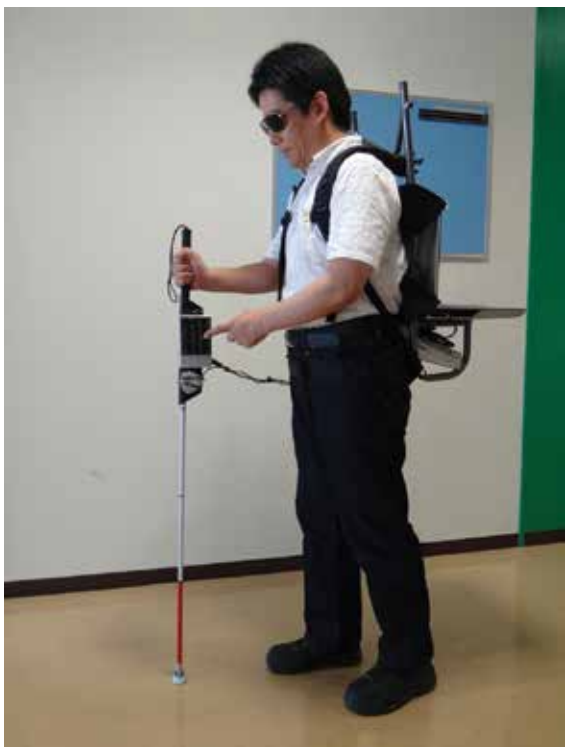

Figure 2. Our Kinect cane system.

the property of a Kinect sensor. These two methods are simultaneously executed in the obstacle detection mode of the Kinect cane system, and if one of these methods detects obstacles, the tactile device returns vibration feedback to the user.

2.1.1. Detection of small obstacles

Figure 3(a) shows an image of a corridor scene including a small obstacle (i.e., a box with a height of 7 cm) on a floor. **Figure 3(b)** shows the depth data of the corridor scene. The scene

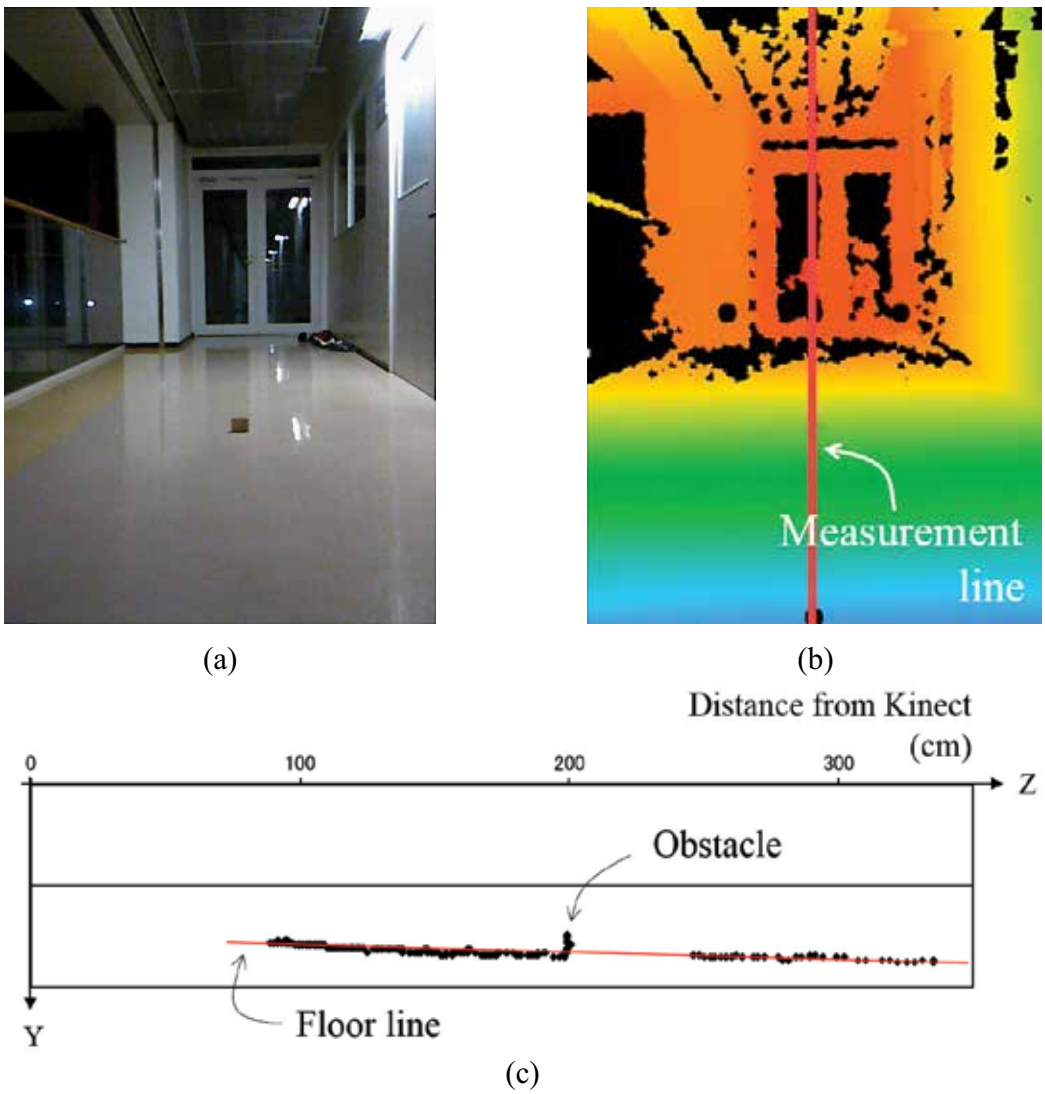

Figure 3. A small obstacle on a floor in a corridor scene. (a) Color image; (b) depth data; (c) profile.

image and depth data were obtained by the Kinect sensor. In the depth data, the distances from the Kinect sensor to points on object surfaces are coded. Black pixels represent positions where the sensor cannot measure the distances. The colored pixels (except black ones) are called *edges* in this chapter.

In order to detect a small obstacle on a floor, the method uses a *measurement line*, which is a vertical line at the center of depth data as shown in **Figure 3(b)**. The measurement line is projected onto the floor plane, and the projected line is called a *floor line*. **Figure 3(c)** shows the profile of the depth data along the floor line. Black dots represent edges on the Z-Y plane.

The floor line is formulated by

$$y = az + b, \tag{1}$$

where a and b are coefficients. Let (z_i, y_i) denote the coordinate of the i-th edge ($i = \{1, 2, \cdots, I\}$). The line is fitted to the edges by minimizing the following sum of the squared distances:

$$S = \sum_{i=1}^{I} (d_i)^2, \tag{2}$$

where

$$d_i = az_i + b - y_i. \tag{3}$$

The optimal values of a and b are obtained by using a robust estimation [69] as follows:

$$\begin{pmatrix} a^* \\ b^* \end{pmatrix} = \begin{pmatrix} \sum w_i z_i^2 & \sum w_i z_i \\ \sum w_i z_i & w_i \end{pmatrix} \begin{pmatrix} \sum w_i z_i y_i \\ \sum w_i y_i \end{pmatrix}, \tag{4}$$

where weight values are defined as

$$w_i = \frac{d_i}{1 + \frac{1}{2} d_i^2}. \tag{5}$$

The equation

$$y = a^* z + b^* \tag{6}$$

represents the optimal floor line. The edges above the optimal floor line are determined to be the obstacle.

2.1.2. Detection of large obstacles

The detectable range of a Kinect sensor is from approximately 40 to 600 cm. In many cases, the upper limitation, 600 cm, is sufficient for obstacle detection. However, the lower limitation, 40 cm, may cause problems that visually impaired individuals collide with obstacles in front of

them. In this section, we propose a detection method of large obstacles not only in the detectable ranges but also nearer than the lower limitation.

In the method, three small circular windows, called *obstacle measurement (OM) spots*, are set on depth data as shown in **Figure 4**. They are arranged horizontally with a certain interval. The positions and interval are determined considering the height and width of the body of a user. Another small circular window, called a *floor measurement (FM) spot*, is set on the bottom area. The OM and FM spots are represented as S_{O1}, S_{O2}, S_{O3}, and S_F, respectively.

If each measurement spot includes enough number of edges, the spot is defined to be *detected*, and the mean depth value is calculated from the depth values of the edges in the detected spot.

The system determines the distance between a Kinect sensor and a large obstacle on the basis of their relation as follows (see **Figure 5**):

Case 1: If the obstacle is nearer than 40 cm, all the spots would not be detected. In this case, the system determines the distance to be less than 40 cm.

Case 2: If the obstacle is between 40 and 600 cm, at least one of the OM spots would be detected, and the FM spot would be detected as well. The system outputs the minimum value among the mean depth values of the detected OM spots.

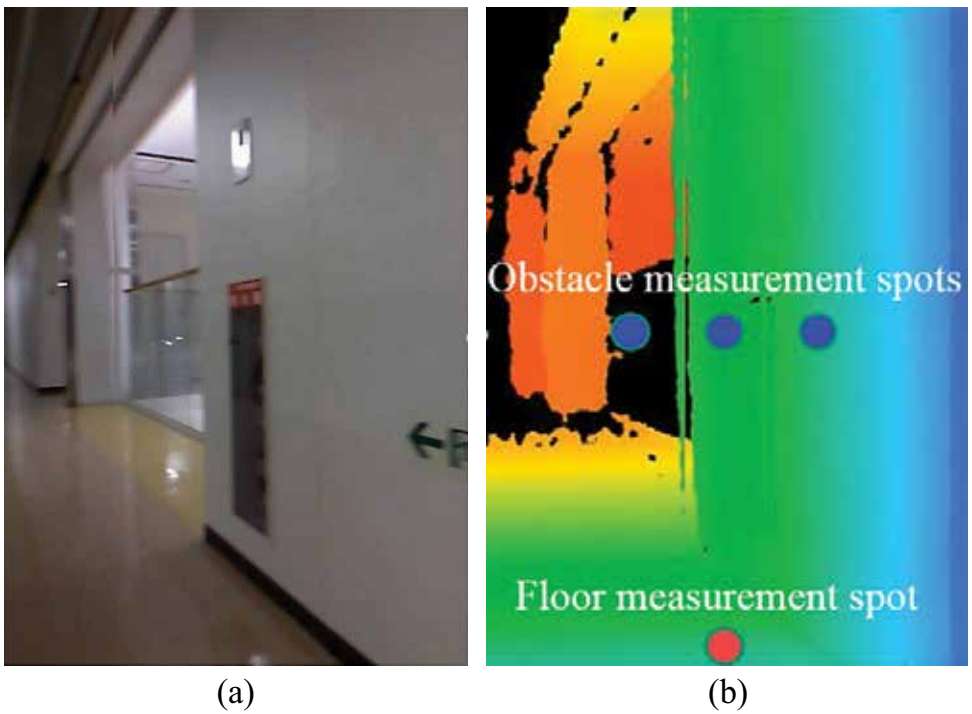

Figure 4. A large obstacle (pillar) in a building. (a) Color image; (b) depth data.

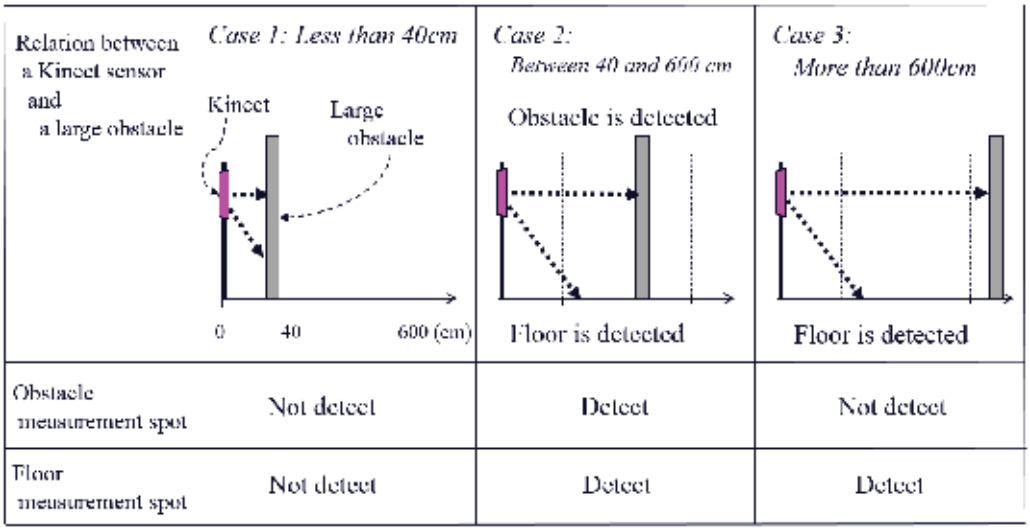

Figure 5. Relation between a Kinect sensor and a large obstacle.

Case 3: If the obstacle is further than 600 cm, all the OM spots would not be detected, whereas the FM spot would be detected. The system determines the distance to be more than 600 cm.

The pillar was successfully detected in **Figure 4**.

2.2. Object recognition

The Kinect cane system can recognize several objects from depth data. The recognition methods and results are described below.

2.2.1. Planes

Artificial environments are generally composed of many planes, such as floors and walls, and therefore, planes can be effective clues to recognize the environments. **Figure 6(a)** and **(b)** shows an example scene and its depth data, respectively. Planes are recognized [70] by using the following method based on random sample consensus (RANSAC) algorithm [71]:

1. Three edges are randomly chosen from edges in depth data, and then a plane is fit to the chosen edges by use of the least-square method. The three pink points in **Figure 6(c)** and **(d)** are the randomly-chosen edges, and the blue regions in **Figure 6(e)** and **(f)** are the planes.
2. The method determines edges whose distances to the plane are nearer than a threshold. These edges are called *inliers*.
3. Steps (1) and (2) are iterated a certain number of times.
4. The plane with the most inliers is selected. **Figure 6(g)** and **(h)** shows the selected plane, which corresponds to the floor. The inlier edges are eliminated.

5. Steps from (1) to (4) are iterated until the number of the remaining edges is less than a threshold.

In **Figure 7**, the floor and the two walls were recognized correctly. Black pixels are the remaining edges, which are used for seat recognition described below.

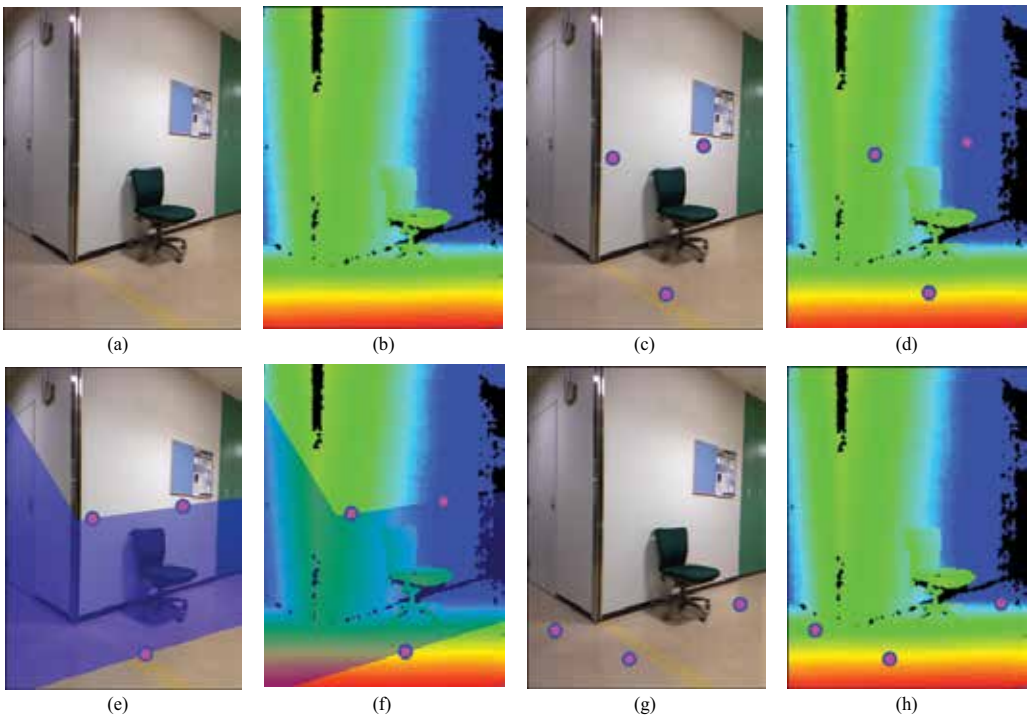

Figure 6. Processes of recognition of planes (a)-(h).

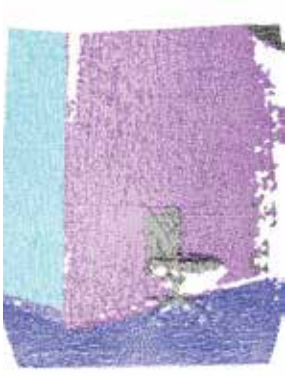

Figure 7. Recognition result of planes.

2.2.2. Seats

The sitting surfaces of seats (such as chairs, stools, and benches) are considered to be the most essential parts of the seats and are recognized as regions that satisfy the following conditions:

1. Candidate regions are composed of the remaining edges which are between 30 and 50 cm from the floor.
2. The areas of the candidate regions are more than 1200 cm ^{2}.

Figure 8 shows the recognition result of the seat in **Figure 6(a)**. Red pixels represent the sitting surface of the seat. **Figure 9** shows the color image, depth data, and recognition result of other seats. All the seats were recognized correctly.

Figure 8. Recognition result of a seat.

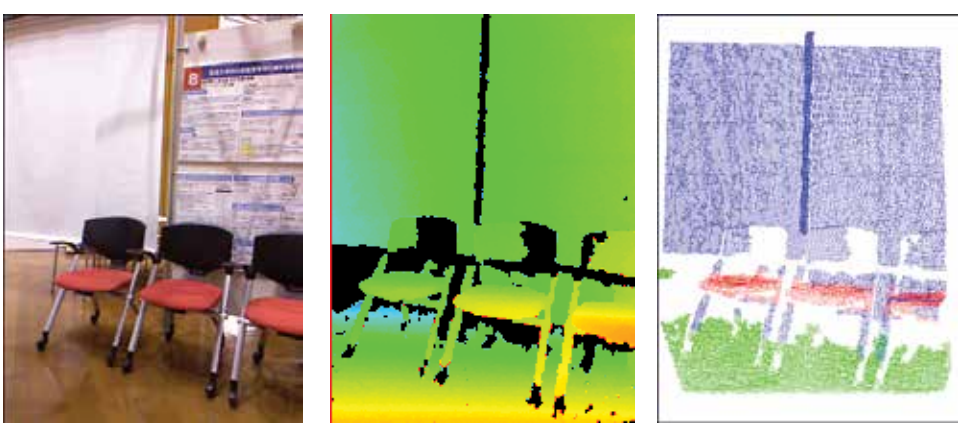

Figure 9. Recognition result of other seats. (a) Color image; (b) depth data; (c) recognition result.

The seat recognition method was applied to 62 sample scenes including seats, and 88% of the scenes were recognized correctly.

It is difficult for this method to recognize seats that have nonparallel sitting surfaces, but there would be not so many such seats in general environment.

2.2.3. Other objects

The Kinect cane system can also recognize upward staircases, downward staircases, and elevators on the basis of the recognition results of planes. The recognition methods are described in detail, for example, in [68, 70], and, in this section, the recognition results are shown in **Figures 10–12**.

It is difficult to recognize upward staircases composed of only one or two steps, slopes, and elevators with sealed doors. The Kinect cane system is not designed to detect holes. A user can detect holes by using the system as a conventional white cane.

2.3. User interaction

Ordinarily, a visually impaired user can use the Kinect cane system as a conventional white cane. **Figure 13(a)** shows an example situation where a user walks in an elevator hall. The user has been here several times, and therefore the user knows there is a bench in the hall, but does not know (or forgot) its accurate location. The user stops walking for safety and then instructs the system to find the bench (seat) as shown in **Figure 13(b)**. The user executes the seat

Figure 10. Recognition result of upward staircases. (a) Color image; (b) recognition result.

Figure 11. Recognition result of downward staircases. (a) Color image; (b) recognition result.

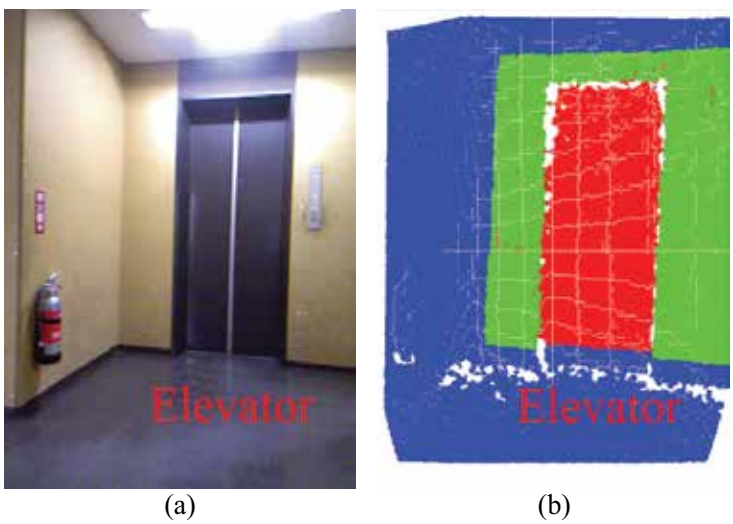

Figure 12. Recognition result of an elevator. (a) Color image; (b) recognition result.

recognition program by pushing the corresponding key on the numeric keypad. The user makes the system search for the bench (**Figure 13(c)**). If the sensor finds the bench, the tactile device returns vibration feedback to the user (**Figure 13(d)**). The user walks toward the bench (**Figure 13(e)**), and then confirms it (**Figure 13(f)**). Finally, the user can sit on the bench (**Figure 13(g)**).

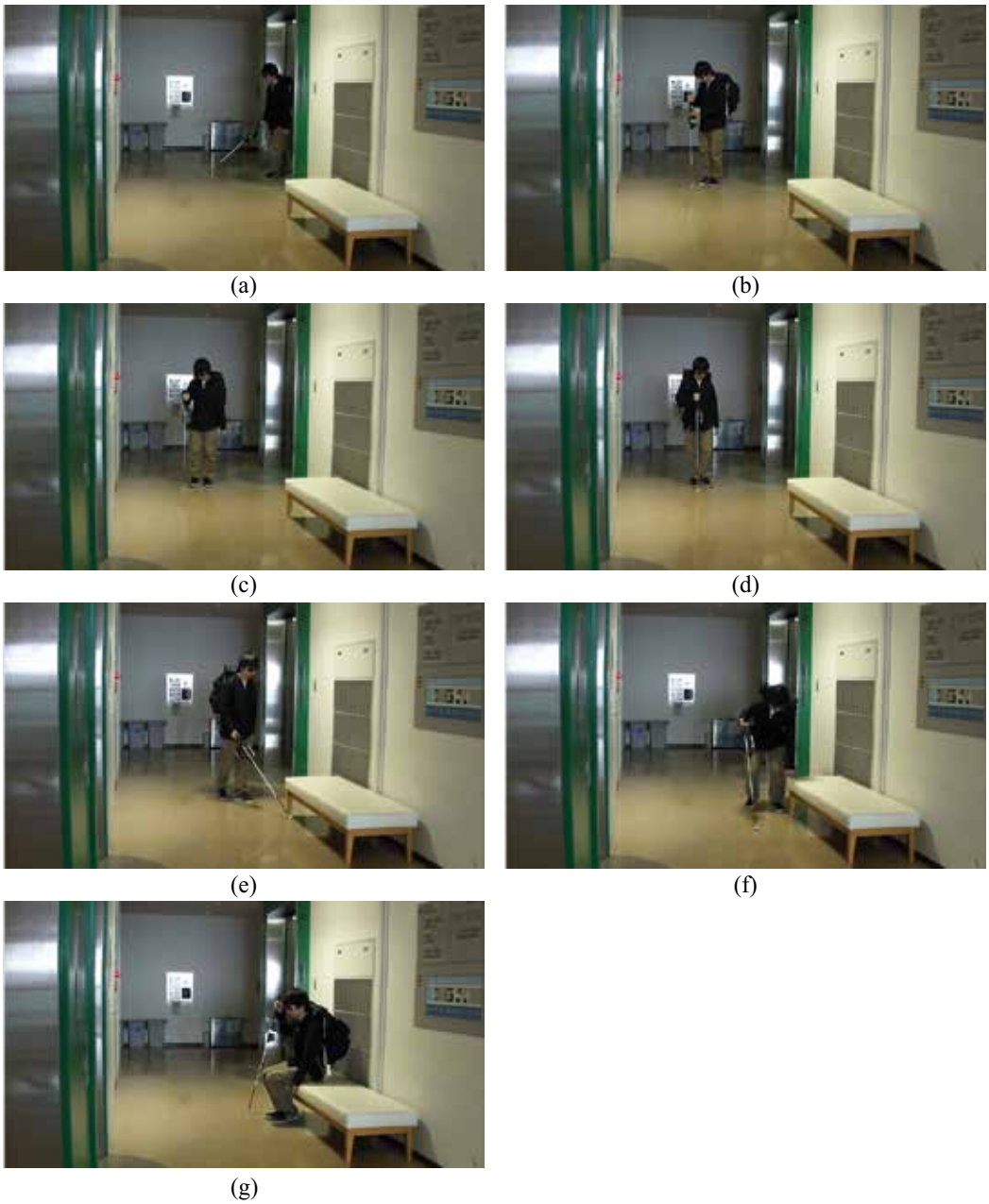

Figure 13. A visually impaired user wants to sit on a bench to take a rest in an elevator hall. (a) A visually impaired user comes out of an elevator. (b) The user instructs the system to find a bench. (c) The user pans the Kinect sensor. (d) The system finds the bench. (e) The user walks toward the bench. (f) The user confirms the bench. (g) The user can sit on the bench.

3. Kinect goggle system

This section introduces another type of Kinect-based wearable assistive system, a Kinect goggle system [72] (**Figure 14**). A Kinect sensor is attached to a goggle on the face of a visually impaired user. A notebook computer, a numeric keypad, a tactile device, and a UPS battery are set in a shoulder bag. These devices are connected with wires for device controlling and power supply. The Kinect goggle system does not require a visually impaired user to hold a heavy Kinect sensor, and therefore the user can make his or her hands free.

The current system can detect obstacles and recognize seats by use of the software which is almost the same as those of the Kinect cane system. **Figure 15** shows an example scene including a bench. The red region in **Figure 15(c)** represents the sitting surface of the bench.

Figure 14. Our Kinect goggle system.

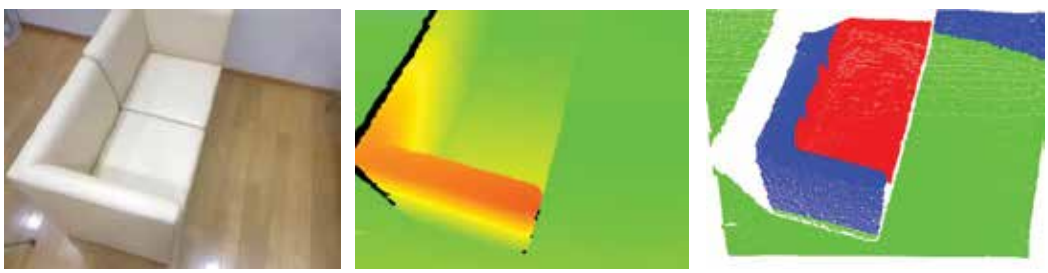

Figure 15. Recognition result of a seat by the Kinect goggle system. (a) A bench; (b) depth data; (c) recognition result.

4. Smartphone-based light checking system

Currently, many visually impaired individuals become to use smartphones as well as sighted people, and therefore it is useful to build assistive systems based on smartphones. In this section, we propose a notification system of the ON/OFF states of room lights and elevator button lights by use of a smartphone camera [73].

4.1. Room light

It is said that many visually impaired individuals often use room lights for the prevention of crime, and so on. They cannot confirm (or have difficulties in confirming) the ON/OFF states by seeing the lights, but can change the states by using the switches of the lights.

There are two types of room light switch. Ones are switches that change their shapes according to the ON/OFF states. The others are switches that do not change their shapes. **Figures 16** and **17** show examples of the former and latter switches, respectively.

Visually impaired individuals can confirm the ON/OFF states if the switches are the former types, but cannot if the latter. It is necessary to help visually impaired individuals know whether room lights are ON or OFF. This section proposes a notification system of the ON/OFF states of room lights based on interaction between a visually impaired user, a smartphone, a room light, and its switch.

First, a visually impaired user turns a smartphone camera toward a room light and then changes the switch of the light as shown in **Figure 18**. The system determines the luminosity

Figure 16. Seesaw-type switch. (a) ON; (b) OFF.

Figure 17. Push-type switch. (a) ON; (b) OFF.

change of the light by analyzing the intensity histogram and the exchangeable image file format (EXIF) information of an image obtained by the camera. If the light becomes brighter and then darker, the smartphone system will tell the user that the room light turns ON and OFF, respectively. If the user fails to set the smartphone camera, the system returns nothing. Therefore, the user can be aware of the failure and do again after resetting the camera.

In a lecture room shown in **Figure 19**, a user used the proposed system and was able to correctly determine that the room lights were turned off.

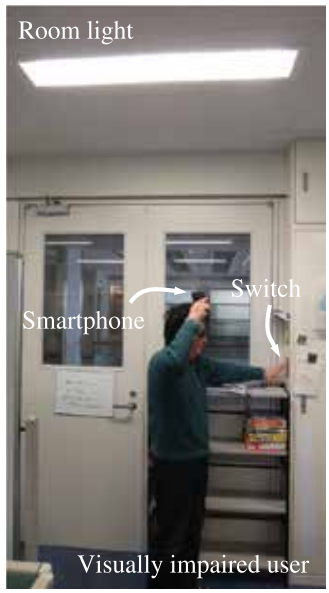

Figure 18. A visually impaired user tries to confirm the ON/OFF state of a room light.

Figure 19. Recognition result of the state of room lights in a lecture room. (a) ON; (b) OFF.

4.2. Elevator button light

The notification system can be also used to confirm the arrival floor of an elevator cage. The light of a floor button on a control panel in an elevator cage often turns off when the cage arrives at the corresponding floor. By using the system, a visually impaired user can know whether the cage arrives at a desired floor as described below.

First, a visually impaired user sets a smartphone camera toward the button of a desired floor as shown in **Figure 20** and executes the light notification program. When the cage arrives at the floor, the light of the button will be turned off, and the system tells the user that the light is turned off. By hearing the message, the user can know the current floor.

In **Figure 21**, a user can correctly determine that he arrived at the ninth floor.

Figure 20. A visually impaired user tries to confirm the current floor.

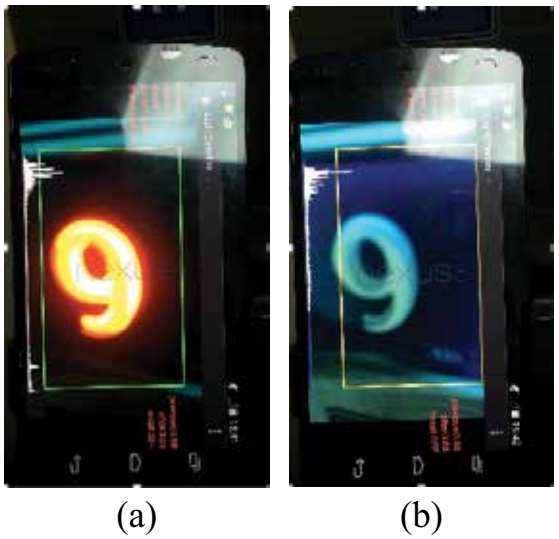

Figure 21. Recognition result of the state of an elevator button light. (a) ON; (b) OFF.

5. Conclusion

In this chapter, we proposed three assistive systems for visually impaired individuals based on image processing. The Kinect cane system can detect obstacles of various sizes and also recognize objects such as floors, walls, seats, upward staircases, downward staircases, and elevators. The detection and recognition results are notified to a visually impaired user by means of vibration feedback. The Kinect goggle system is another type of wearable system and can make user's hands free. The system can also detect obstacles and recognize objects. The smartphone-based light checking system can inform a visually impaired user about the ON/OFF states of room lights and elevator button lights. The user can confirm light states and arrival floors. The experimental results demonstrate that the proposed systems are effective in helping visually impaired individuals in everyday environments.

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Chapter 8

Designing Hands-On Robotics Courses for Students with Visual Impairment or Blindness

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Frank Hees and Sabina Jeschke

Additional information is available at the end of the chapter

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Abstract

School laboratories let students playfully experience the fundamentals of, for example, robotics, computer science, and technology-related topics. By working with LEGO Mindstorms, secondary school students get a chance to learn on a cognitive, emotional, and haptic level and gain experiences with the aid of even more advanced robotics. However, due to an impairment or lack of sight, it is hardly possible for some students to fully participate in a programming process or in building a robot. To overcome this unintentional discrimination, the interdisciplinary student laboratory "RoboScope" at RWTH Aachen University has teamed up with a group of experts to develop a barrier-free robotic course. Since then, the course has been tested and implemented based on concurrent evaluations and frequently held at RWTH and several other German schools. The presented work covers an overview of different kinds of visual impairment and lab settings and the development cycle of the courses at RWTH from design to testing, implementation, and further development regarding the evaluations. Evaluations show that students who are visually impaired or blind appreciate the opportunity to participate in the field of robotics. An insight into the evaluation concept that differs from "regular" courses in the "Roboscope," as well as the results are used for further development.

Keywords: visually impaired, blind, robotics courses, school labs, extracurricular robotics lab

1. Introduction

By introducing students to the fundamentals of robotics in an informal, playful setting, extracurricular school laboratories are an effective way to encourage interest in computer science

or other technology-related topics. In working with LEGO Mindstorms, a set of soft- and hardware to build programmable robots, they get a chance to learn on a cognitive, emotional, and haptic level. Unfortunately, not every pupil is able to participate in courses like these due to a lack of accessibility. For example, Ludi states that "awareness of potential career paths and access to adequate preparation remain barriers to students who are visually impaired" [1]. For pupils who are visually impaired or blind, it is essentially impossible to fully participate in a programming process or in building a robot.

The Center for Learning and Knowledge Management, in conjunction with the Institute of Information Management in Mechanical Engineering of RWTH Aachen University and a group of experts, set out to develop a special and accessible course for the visually impaired and blind students. The experts consulted a group of psychologists, school and university teachers, and experts in the field of accessibility as well as in robotics. They took the original course design from an existing robotics course for high school students and transformed it into an accessible course design. The original course consists of theoretical input about building robots and programming as well as the subsequent practical phases, in which the students apply their knowledge on EV3 roboters. At the beginning of each course, the robotic equipment is explained and the problem that needs to be solved is presented. The students—in compliance with the supervisors—then analyze the problem and identify the necessary steps to be taken and in doing so, the desired outcomes of the experiment are met. The applied combination of both theoretical and practical factors has proven to facilitate an authentic learning environment and strong learning results [2]. However, developing a new and adequate course simply by applying technical adjustments is not sufficient. Therefore, all changes applied to the course went hand in hand with an adjustment of teaching and learning strategies.

When designing a programming course for pupils with disabilities, it is crucial to develop these strategies as well as a list of the required tools as a first step. From the gathered findings, the resulting new course design allows students who are visually impaired to participate in the same courses and benefit from the same experiences—such as programming and building a robot—as their fellow pupils. This paper will present an overview of types of visual impairment, different lab settings and an insight into the original course design. This will be followed by results from the expert design workshops in terms of technical and didactic adjustments to the course. Finally, the paper will conclude with the full development cycle of the courses at RWTH Aachen University from design to testing, implementation, and further development.

2. Visual impairment

Vision, as one of our five senses, enables us to learn about our environment. Being able to see not only helps us to orientate ourselves but also shapes our perception of the world and all it has to offer. Not everyone, however, possesses full vision. "Many people have some type of visual problem at some point in their lives" [3]. Some minor problems can occur, for

instance: in seeing objects that are far away (near-sightedness) or that are extremely close or in very small print (far-sightedness). "These types of conditions are often easily treated with eyeglasses or contact lenses" [3].

According to the World Health Organization (WHO), apart from these minor eye problems, "285 million people are estimated to be visually impaired worldwide: 39 million are blind and 246 have low vision" [4]. Other key facts from the WHO about visual impairment state that around "90% of the world's visually impaired live in low-income settings," "82% of people living with blindness are aged 50 and above" and that 80% of all visual impairment can be prevented or cured [4]. Many people worldwide are only visually impaired because they do not have access to reading aids or medical care in general; most people with visual impairment come from low-income or developing countries. "Globally, uncorrected refractive errors are the main cause of moderate and severe visual impairment; cataracts remain the leading cause of blindness in middle- and low-income countries" [4]. The risk of going blind is estimated to be 10 times higher in developing countries than in industrialized countries [5]. In comparison to persons with low vision, persons who suffer from blindness face additional challenges. These can manifest themselves in social challenges by the difficulty of participation in social activities and events, navigation and orientation in unfamiliar environments as well as difficulties in using technology such as computers and smartphones. Although many people with blindness use tools such as blind rods, screen readers, and other helpful measures, not everyone has the financial background or even enough self-esteem to counterbalance the previously mentioned disadvantages.

According to the National Eye Institute in the United States, "less familiar visual impairments include:

- **strabismus**, where the eyes look in different directions and do not focus simultaneously on a single point;
- **congenital cataracts**, where the lens of the eye is cloudy;
- **retinopathy of prematurity**, which may occur in premature babies when the light-sensitive retina has not developed sufficiently before birth;
- **retinitis pigmentosa**, a rare inherited disease that slowly destroys the retina (see **Figure 1**);
- **coloboma**, where a portion of the structure of the eye is missing;
- **optic nerve hypoplasia**, which is caused by underdeveloped fibers in the optic nerve and which affects depth perception, sensitivity to light, and acuity of vision; and
- **cortical visual impairment** (CVI), which is caused by damage to the part of the brain related to vision, not to the eyes themselves" [4].

In contrast, there are no reliable numbers of visually impaired children and teenagers in Germany. Nevertheless, visual impairment is a big issue and causes imbalances, particularly

Figure 1. Retinitis pigmentosa, http://www.rsb.org.au/retinitis-pigmentosa.

in education, higher education and career opportunities, especially in Science, Technology, Engineering and Mathematics (STEM) focused education. The robotics courses for students with visual impairment or blindness at RWTH Aachen therefore aim to overcome these inequalities.

3. Robotics laboratories

3.1. Student laboratories

The current technological developments being triggered by Industry 4.0—the combination of industrial production with modern means of communication—as well as digitalization in general pose major challenges for robotic education; hence, the demand for students from fields affiliated with science, technology, engineering, mathematics, and robotics in particular is steadily increasing. Due to concern related to this increasing demand for future engineers and the significance of qualitative scientific and technological education, universities and other seats of learning are focusing on secondary school students. Hence, since 1990, school labs have been an important component of German universities [6, 7].

Student laboratories are extracurricular educational institutions, which allow children, pupils, and students to experience science. New devices and technology can be used and tested for clear understanding and unconventional learning of modern research techniques. These laboratories often focus on natural science fields and foster insight from range of topics in a field of study. Extracurricular learning venues of universities, which are concerned with robotic science are often available exclusively for students and scientists. Furthermore, many schools do not have the resources to purchase devices and equipment in order to implement laboratories in class. The school laboratories of RWTH Aachen University, in contrast, focus on those pupils and younger students and enable them to discover distinct capabilities while learning by testing and playing.

3.2. Student laboratories in Germany

Around the globe, there are laboratories available that also work in equal or related fields of robotics, though very few are constantly available for secondary school students. This is largely due to the fact that equipment is often very expensive, difficult to acquire, and too conceptually complex for young persons to use. The special challenge of the RoboScope (http://www.robo-scope.de/home.html) of RWTH Aachen University is to give students—both sighted as well as visually impaired or blind—something that inspires them and makes them curious. By giving them an achievable, yet challenging task, it aims to foster a desire to work in a respective field of engineering. In the following, a few examples of robotics labs for students in different parts of Germany will be presented.

Other universities like the Technical University of Hamburg/Harburg cooperate with companies to encourage pupils to learn programming skills. For instance, they offer seven different courses based on interests and experiences at the university, which may consist of weekly meetings at the school or participation in a voluntary project team. In different modules, such as a trial course, pupils learn while using LEGO Mindstorms robots and basic graphic programming. In higher modules, they get introduced to programming languages like C and C++, soldering and building a LEGO-Mindstorm robot [8].

The concept of offering certain courses based on a student's interests and experiences is also common across other universities or institutions. The Technical University of Kaiserlautern provides three different classes, from basic programming to getting a robot to follow lines in a labyrinth up to a course in preparation for a robotics tournament. The main field of attention lies in sensor technology with the aid of tactile, light, and ultrasonic sensors [9].

The "TUMLab," the Technical University of Munich's lab situated in the German Museum, offers a similar course based on this technology. In five different modules, pupils get to know diverse sensors and how to apply them ingeniously. Herein, lies the main goal of getting a robot to find its way around autonomously [10].

The "Technikum29" in Kelkheim-Hornau in Germany offers a workshop for learning about and using sensors, branches, subprograms, busses, interrupts, and how to develop logical decisions and games using a Raspberry Pi and similar single-board computers. As a distinguishing factor from the robotics summer camp of the University of Darmstadt, which focuses on ages 10–14, the Technikum29 requires its participants to be at least 14 years old. At the robotics summer camp, younger pupils learn to communicate using Bluetooth technologies and to build their own robot with a LEGO Mindstorms packet. Older pupils get to discover and solve problems and tasks given from the instructors, who are computer science students at the University of Darmstadt. Both courses take place during 1 week of the summer holiday and have included children with disabilities since 2013 [11].

3.3. Student laboratories worldwide

In Switzerland, the ETH Zürich focuses on preschool children and offers a "Bee-Bot" kit that consists of a child-friendly bee-shaped robot. Teachers can rent six small robots, playing cards, and teaching accessories such as activity mats and charging stations for 2 weeks. The

small robots are programmable with four buttons and can be moved over a map easily. Before renting those sets, teachers get a short workshop at ETH where they learn about basic robotic science.

Elementary schools can participate in a similar project. Teachers are trained by a research team from ETH, who also give advice and support while using the technology in the classroom. Topics of this project are the concept of computational thinking and the functionality of robots. Lessons are arranged as project-based learning, and pupils learn to program robots playfully.

A different project of the ETH is the "RoboMINT" in which children learn to build a robot. In a "Dancebot course," pupils learn to solder and program a dance-choreography for the robot they have built. A second course uses small lights attached to a robot and a camera with long time exposure to draw a picture. For the picture, the robot uses a coded paper to follow lines. The sets can be rented for free [12].

The DNA Learning Center (DNALC), which is promoted by the United States of America's Cold Spring Harbor Laboratory, a private, not-for-profit research and education institution at the forefront of molecular biology and genetics, offers class field trips and summer schools devoted entirely to public genetics education [13].

3.4. Student laboratories for disabled children

Besides RWTH Aachen University and the Technikum29, the Bayer Science & Education Foundation has provided Anna-Freud-School in Cologne with an accessible laboratory for pupils. Laboratory equipment and computerized workplaces were purchased with the budget of 22,000 Euros. These new features allow children with disabilities to work on projects independently and to identify and nurture talents early. In this way, the school is able to promote pupils who are physically disabled or have a chronic or psychosomatic illness. The Foundation supports projects, which are used to complement lessons in school and to draw interest in natural science and technology [14].

The Perkins School for the Blind in Massachusetts (USA) offers short courses in robotics for secondary school students (Grades 6–10), who can learn about the highly sought-after skills of mechanical and electrical engineering, computers, math, and science. Participants work with their peers and knowledgeable staff to build basic robots and program them to complete simple tasks. Inconveniently, though, this course is currently not offered on a regular basis.

Learning about robotics is an enjoyable and exciting way for students to increase knowledge in the areas of science, mathematics, and technology and provide students with an opportunity to gain first insights. Our extensive research has shown that around the globe, there are laboratories available that also work in equal or related fields of robotics, though very few are both constantly available for students and offering a chance at hands-on experience.

To foster an interest in STEM-fields, it is necessary to involve pupils in the process of programming a robot playfully. Technical universities in Germany and Switzerland already offer a broad range of courses for pupils to learn basic programming starting at a young age.

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In recent years, reform efforts in science curriculum have stressed the integration of educational technology into teaching and learning purposes. Teachers and educators face the challenge and the chance to explore inventive ways in which new technologies can be utilized to improve accessibility to science for students with visual impairments. Integrating students with disabilities in student laboratories is an effort that not only encourages interest in STEM-fields but also shows these individuals opportunities for their future careers. Technological resources for people who are visually impaired, like Braille generating software, Braille printers, screen-reader software, and speech synthesizers, already exist.

Teachers, educators, and educational institutions need to realize the student-oriented benefits and put more effort into accommodating students with (visual) impairments in STEM education. An awareness, and furthermore, an understanding of the academic needs of students with (visual) impairments are essential in striving toward this goal. Unless many institutions and educators stress the need to integrate students with (visual) impairments in their scientific programs, there is still a long way to go. Fully accessible participation in science will be beneficial for all students and a rewarding experience for teachers.

4. Original course design: "Roborescue" and "Rattlesnake"

At RWTH Aachen University, high school students are given the chance to gain insight by using LEGO Mindstorms construction sets in a school laboratory and constructing and programming various robot models. They are using the graphical programming interface NXT-G to discover an easy introduction to programming, since it is suitable for nonprofessionals [15].

To prepare and motivate students for a future career in robotics, the course program allows students to try their hand at building, programming, and testing robots in a highly interactive and playful environment. In order to captivate students, the course allows them to create either a "rescue robot" [16] that can search for virtual victims in a simulated rescue mission or a "rattlesnake" that snaps shut when someone crosses its field of vision. The choice of the scenario is subject to the age of the students—lower grades create a rattlesnake, (which is easier to build and to program) while junior and senior classes go on a more complex rescue mission. The four main phases of the course are: the introduction, which gives basic information; the construction; the programming process; and the reflection or evaluation phase. To follow along a learning process, the underlying didactic course concept focuses on individual practical, experimental, and playful experiences [15]. In accordance with the feedback of the course participants, this course design was chosen to build up an extracurricular learning venue for students with visual impairment and blindness to give them first insights into robotics.

The educational laboratory is not located at the students' respective schools; rather, it has been set up at RWTH Aachen University. This allows high school students to take a peek into the daily routine at university and is also meant to facilitate the decision-making process when it comes to choosing further steps after graduating from high school [17].

5. Enabling higher accessibility for visually impaired students

5.1. Expert design workshops

To facilitate the process of redesigning the robotics course in order to reach a higher level of accessibility for students with visual impairment as well as blindness, researchers from RWTH Aachen University invited a team of interdisciplinary experts to a series of workshops. The roadmap of the redesign was developed within these workshops. The main goal was defined as follows: to identify the key aspects of required adjustments.

During the workshops, the participants gradually developed a grid of these requirements. In a first step, they divided the course into its individual phases based on the established approach by Vieritz et al. [18]. They used the different phases of the course and analyzed the requirements and necessary adjustments for each individual part compared to those of the original course design. These phases consist of the introductory part, the construction phase, the programming phase and the phase for reflection. Combining their different experiences and testing single elements by simulating specific eye dysfunctions, the experts came to results in terms of requirements for each phase. These results are presented and discussed in the chapters below, which are divided into technical as well as didactic adjustments. At the end of chapter three, the developed grid gives a summarized overview of the results from the workshops.

5.2. Technical requirements

According to the results of the design workshops, the identified requirements especially include auxiliary means, which can be summed up as objects, software, and computer settings. There are a lot of different eye dysfunctions which call for support by varying objects, for example, magnifiers and common magnifying glasses. Other important objects for different phases of the course are cameras and reading devices, printed handouts for every phase, additional lighting for the building process, and sorting boxes for robot components.

In terms of software, screen readers such as JAWS or Dolphin, graphic programming using, for example, NXT-G [14] as well as textual programming using, for example, JBrick [19, 20] should be provided in the programming phase. Additionally, the computers and provided worktables should allow adjustments of graphic contrast on computer screens. Nevertheless, there is no “universal remedy” for increasing accessibility. In preparation of the course, the teaching staff needs to communicate with the participants to be prepared for any special requirements the students might have.

5.3. Didactic adjustments

Since not every measure taken is helpful for every sort of handicap and not all changes can be made at once, it is necessary to differentiate between the types of visual impairment. In the presented case, a fundamental distinction between different degrees of visual impairment up to sightlessness has been the essential groundwork for further research and course

development. To reach full accessibility for the pupils, advancements and changes must be made gradually. This methodology has proven to be a very helpful approach in the process of designing the new course. Some degrees of visual impairment, for example, are even contrary to one another [1], so there is an increasing demand for different technical as well as didactic approaches in each course to reduce or extinguish existing barriers for all participating students.

As a first result and requirement, printed manuals with regard to font size should be provided within the first three phases: the introductory part, the construction, and the programming phase. This allows students with less severe visual impairment to be able to reread instructions at their individual pace.

Time has also proven to be one major but often underestimated factor [21]. Students who have visual impairments need to be given more time to work on their tasks in terms of reading instructions, following presentations as well as building and programming. The more severe the impairment, the more time will be needed to finish a task. Kabátová et al. [21] found that test participants who were wearing glasses that simulate an eye dysfunction needed four times as long to finish the assigned task without the glasses. Therefore, they come to the subjective conclusion that the time necessary for a traditional course design should be multiplied by a factor of at least four. Further research and evaluations of the course will have to prove whether that factor needs to be adjusted.

Another important adjustment relates to the teacher-student ratio. It has to be increased compared to traditional course designs, which of course takes up additional time and resources on the teaching end. The required ratio can differ vastly, as students have very diverse needs in terms of support. As we also know from Silva et al., even students without handicap perceive and process experiences in different preferred ways [22]. As a result, the instructors need to provide a high level of flexibility regarding supervision and must provide support throughout the course. Lastly, the supervisors identified pre-sorting the sorting boxes used in the construction phase as a helpful measure in the building process, which no longer excludes students with visual impairments from the haptic and tangible experience of building a robot themselves. Every course is highly influenced by diverse aspects, and a thorough preparation and awareness of all possibilities and influences as well as a pre-analysis of the expected target group of each course proves to be the key to a successful course design. **Figure 2** sums up the results from the workshop in a grid.

5.4. Further development of the courses

After an implementation of the workshop and the guidelines, robotics courses were conducted in cooperation with the Berufsförderungswerk Soest and teenagers of the Johannes-Kepler-School in Laurensberg. An excursion into the mode of operation and programming of industrial robots was made for the course's participants.

The robotics courses for visually impaired teenagers were perceived very well and were therefore asked to be offered to young people who are blind. Since the robots could not be programmed with the LabView-based programming language NXT-G, a different setup was required for the course. The "Blindenstudienanstalt Marburg" was visited for preparation

Phase	Content	Original Course Design and equipment	Technical Requirements for a barrier free course	Didactical Requirements for a barrier free course
Introduction	Theoretical Input	Power Point Presentation	- Laptops with screen readers - Magnifying glasses	- Detailed explanations and descriptions of what the slide shows - Repetition of content - Simple slide design with high contrast - Printed Manuals
Construction	Building of the robot	Unsorted boxes	- Sorting boxes - Magnifying glasses - Reading Device - Graphic contrast on work tables	- Pre-sorting of components - Room for extra time and practice - Continuous supervision and support - Printed construction manuals
Programming	Programming of the robot	Laptops	- Contrast settings - Screenreader (JAWS/Dolphin) - Extra lighting - Printed Manual instead of beamer - On-screen magnifier - Graphic programming	- Continuous supervision and support - Room for extra time and practice - Printed programming manuals
Reflexion	Reflecting the Processes and Outcomes			Room for extra time

Figure 2. Results from the workshop: requirements for the new course design.

in June 2014. Since the sense of touch system is the most important part for people who are blind, the "burrowing" in Lego boxes and the "building" of the robots are the most important components of the course regarding blind-pedagogical aspects. On top, a spoken construction guide is provided to the students as an audio book. The written construction manual was examined by the teachers of the "Blindenstudienanstalt Marburg" and was then tested in the course. **Figures 3–6** show impressions of the prepared soft- and hardware as well as students in the course programming a robot.

In 2015, the robotics courses for students who are blind were implemented in cooperation with the Blindenstudienanstalt Marburg. The building instructions, which were previously unusably for people who are blind, were converted into a spoken building instruction. The

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Figure 3. Prepared Lego Mindstorm system for students who are blind.

Figure 4. Secondary school student who is blind programming a robot.

Figure 5. Prepared software for the robotics course for students who are visually impaired: textual instructions.

instructions themselves were then read out by a free screen-reading program NVDA via a voice output system to create real-world conditions. The program code was read out or put out via a Braille display reader, which allowed even the programming and integration

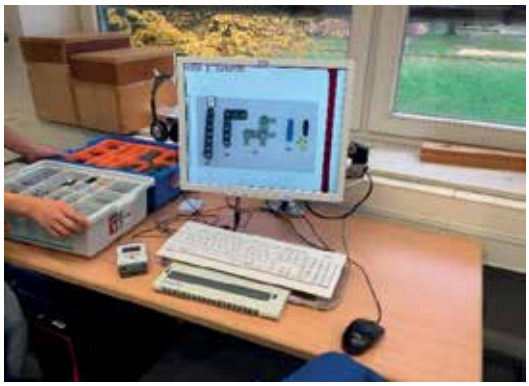

Figure 6. Prepared software for the robotics course for students who are visually impaired: illustrated instructions.

of sensors with conditions, loops, and queries. The robotics courses for pupils with visual impairments are frequently requested by different classes and are met with great enthusiasm. Usually, the course takes place between two and five times a year. Furthermore, teachers can request the developed materials, rent LEGO Mindstorm robots, and ask for advice on how to conduct the courses on their own.

6. Evaluations of the course

The evaluation of the courses took place verbally and was later put onto paper. With the help of statistical evaluation methods, feedback can be recorded quantitatively in order to validate the success of the measure or to adapt it. In addition to inquiries such as "How did you like the lecture/the programming/the difficulty of the tasks/the course schedule?" (6-degree scale from 1 = very good to 6 = not satisfying) or "How did you like the day?" (3-degree scale from 1 = too easy to 3 = too difficult), the questionnaire includes open questions that allow room for comments and suggestions for improvement.

The pupils (n = 8) mentioned, on a 6-degree scale, that the presentation was very exciting (Median = 1.25), that the programming had been very good (M = 1) and that they learned something (M = 1.13). Every pupil also said that they enjoyed the course and that he/she would like to visit the course again. Some of the students mentioned in the open questions section that they wanted a successor program, which will be developed as a consecutive step. Due to the small sample size, the statistical power of the results is reduced and statistical relevance of the quantitative data needs to be discussed. However, from a qualitative point of view with regard to the frequently given feedback, the analysis showed that the students' enthusiasm, interest, and appreciation for the courses were high among both teachers and students.

7. Conclusion and outlook

To keep up the interest in STEM-fields, it is necessary to involve pupils in the process of programming playfully. Technical universities in Germany and Switzerland already offer a

broad range of basic programming courses for pupils starting at young age. Additionally, to encourage applied computer science in school, some institutions offer robot kits and teaching materials. Integrating students with visual impairment or blindness in student laboratories is a chance to not only encourage interest in STEM-fields but also to show these individuals opportunities for the future. Science curriculum reform efforts have emphasized the integration of educational technology into teaching and learning purposes in the past years. Teachers and educators are asked to explore further ways in which new technologies could be utilized to improve access to science for students with visual impairments.

The paper has described the process of redesigning of a robotics course from an educational robotics laboratory to increase accessibility of the course for students with visual impairments. The evaluation of the workshop has informed a concept for the redesign, which has been implemented and is currently being tested in a second run with various groups of students with visual impairments. The developed grid of the workshop suggests that adjustments to the designated phases of the lecture can provide a higher level of accessibility. A first anecdotal but enthusiastic assessment from the students who participated leads to the assumption that the applied suggested changes were successful.

Nevertheless, a huge part of the adjustments requires a consideration for the unique needs and requirements that the specific dysfunctions of the target group bring about. At this point in the research, there is no catch-all solution to the challenge. The evaluation of the designed courses will allow for a thorough analysis, serve the pursuit of continuous improvement, and be the key to future research. Additionally, to broaden the range of accessibility, further research will have to focus on full accessibility not only for those students who are blind but also for those with other impairments, such as hearing and physical disabilities.

In conclusion, teacher, educators, and educational institutions should realize and promote the student-oriented benefits and devote additional effort toward accommodating students with (visual) impairments in STEM education. An awareness, and furthermore, an understanding of the academic needs of students with (visual) impairments are essential in striving toward this goal.

Though many institutions and educators stress the need to integrate students with (visual) impairments in their scientific programs, there is still room for improvement. Ensuring that full participation in science is possible for everyone will be beneficial for all students and a rewarding experience for teachers.

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About 4% of the world population has visual impairment or blindness. This book is aimed at addressing different causes of visual impairment and blindness, their epidemiology, manifestations, risk factors, prevention of progression, and treatment. It is aimed at encouraging physicians and researchers to increase efforts to prevent irreversible and treat reversible blindness for the betterment of the world. Therefore, it is essential to be fully aware and knowledgeable of the manifestations of the diseases causing blindness, and this book covers some of their different aspects. Each chapter was written by experts from around the globe. Thus, it reflects the importance of the subject.

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