

Falls Risk Assessment in People with Retinitis Pigmentosa

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Abstract

Visual impairment is a major risk factor for falls in older adults, but it is not known what specific falls risk factors are present in adults with impaired vision. The purpose of this study is to examine whether standard clinical outcome measures that have been validated to identify falls risk in older adults would adequately identify falls risk in individuals with visual impairment due to retinitis pigmentosa (RP). Ten adults with RP completed clinical gait and balance assessments, as well as visual function testing. The majority of participants had normal scores on gait and balance measures, yet 8 out of 10 had experienced at least one fall within the past year. The relationships between falls history, gait and balance measures, and visual fields were in the opposite direction to predictions. For example, faster gait speed, typically associated with better stability, was correlated with more falls. Further research is warranted in order to better elucidate the specific falls risk factors in individuals with impaired vision so that appropriate interventions can be developed.

Keywords: falls risk, outcome measures, retinitis pigmentosa

Introduction

It is well documented that older adults are at increased risk for falling and experience a greater rate of falls-related injuries. Falls are caused by multiple factors and typically result from the interaction of biological risk factors (e.g., individual factors, such as muscle weakness) and environmental risk factors (e.g., poor lighting). The most important intrinsic risk factors for falls include muscle weakness and gait and balance disorders, together

with visual impairment (American Geriatrics Society, 2001). Visual impairment increases the odds of a fall by approximately 2.5 times (Rubenstein & Josephson, 2006). Various studies have identified specific visual deficits associated with the incidence of increased falls in older adults, including reductions in visual fields, depth perception, and contrast sensitivity (Freeman, Muñoz, Rubin, & West, 2007; Ivers, Cumming, Mitchell, & Attebo, 1998; Lord & Menz, 2000). These visual deficits can result in increased tripping as a result of not seeing hazards, falls on stairs due to poor depth perception, or failure to notice a change in surface due to poor contrast sensitivity.

A major goal in therapy is to identify individuals at risk for future falls in order to intervene before a fall

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occurs. Physical therapists typically use performance-based outcome measures of balance and gait in order to identify those individuals at risk for future falls. Researchers have tested older adults to develop criteria that optimize sensitivity and specificity of each outcome measure in order to predict future falls. One of the simplest measures used to identify older individuals at risk for falls is preferred gait speed. Gait speeds less than 0.7 meters per second have been associated with increased falls incidence (Monterro-Odasso et al., 2005). The Dynamic Gait Index (DGI) assesses an individual's ability to modify gait in the presence of external demands; total scores lower than 20 (out of a possible maximum score of 24) indicate falls risk (Shumway-Cook, Baldwin, Polissar, & Gruber, 1997; Whitney, Hudak, & Marchetti, 2000). The items of the DGI include walking with changing speed or head turns, walking over and around obstacles, and stair climbing. Computerized dynamic posturography assesses the ability to use sensory input to maintain balance. Sensory input is systematically reduced during the Sensory Organization Test (SOT) and total composite scores of less than 38 (out of a possible maximum 100) have been associated with recurrent falls (Whitney, Marchetti, & Schade, 2006).

Visual deficits are associated with postural instability and reduced mobility, and, as a result, increased incidence of falls. For example, a one-line reduction in visual acuity was associated with a 10 percent increase in the odds of mobility restriction, whereas visual field loss of 10 percent was associated with a 20 percent increase in the odds of mobility restriction (West, Gildengorin, Haegerstrom-Portnoy, Schneck, Lott, & Brabyn, 2002). Although it has been argued that peripheral visual field loss is more critical to postural stability and therefore falls incidence, studies have demonstrated that both central and peripheral visual field loss result in increased falls incidence. Older women with central visual field loss due to age-related macular degeneration (AMD) have significantly greater falls risk than their peers as a result of impaired balance, slow visual reaction times, and poor vision (Szabo, Janssen, Khan, Potter, & Lord, 2008). As a result, women with AMD fall at nearly twice the rate of women without AMD (Szabo, Janssen, Khan, Lord, & Potter, 2010). Patients with glaucoma, which results in loss of peripheral vision, have three times the risk

of falls compared to those without glaucoma (Haymes, Leblanc, Nicoleta, Chiasson, & Chauhan, 2007).

Retinitis pigmentosa (RP) is a term applied to a group of hereditary retinal degenerative diseases characterized by the loss of function of the rod and cone photoreceptors. It is estimated to affect approximately 1 in 3,700 people in the United States (Boughman, Conneally, & Nance, 1980). The primary symptoms of RP include night blindness (impaired vision at low light levels due to rod degeneration early in the progression) and constricted peripheral visual fields (Berson, Sandberg, Rosner, Birch, & Hanson, 1985; Feliuss et al., 2002; Grover, Fishman, Anderson, Alexander, & Derlacki, 1997; Massof & Finkelstein, 1981; Woods, Giorgi, Berson, & Peli, 2010). Other symptoms can include reduced visual acuity, reduced contrast sensitivity, and photopsias (Alexander, Derlacki, & Fishman, 1995; Berson, Rosner, Weigel-DiFranco, Dryja, & Sandberg, 2002; Bittner, Diener-West, & Dagnelie, 2009; Holopigian, Greenstein, Seiple, & Carr, 1996). RP can also lead to cataracts at a relatively young age (Jackson, Garway-Heath, Rosen, Bird, & Tuft, 2001), and is known to have an impact on many aspects of everyday function, including face recognition, color perception, and reading (Barnes, De l'Aune, & Schuchard, 2008; Fishman, Young, Vasquez, & Lourenco, 1981; Szlyk, Fishman, Alexander, Revelins, Derlacki, & Anderson, 1997).

The effects of RP on balance and mobility have been investigated in a number of studies using both self-report measures and performance-based measures (Black, Lovie-Kitchin, Woods, Arnold, Byrnes, & Murrish, 1997; Hartong, Jorritsma, Neve, Melis-Dankers, & Kooijman, 2004; Haymes, Guest, Heyes, & Johnston, 1996; Turano, Geruschat, & Stahl, 1998; Turano, Geruschat, Stahl, & Massof, 1999; Turano, Herdman, & Dagnelie, 1993), but the issue of falls risk has not been addressed as much. Studies have demonstrated that individuals with RP often cannot use available visual cues appropriately to stabilize balance and will exhibit increased postural sway (Turano et al., 1993). In addition, individuals with RP often have reduced gait speed and increased accidental contact with objects on obstacle courses in the environment, under both normal and reduced illumination (Black et al., 1997; Geruschat, Turano, & Stahl, 1998). Increased postural sway and reduced

gait speed typically indicate decreased stability, which in turn, increases falls risk.

Thus, the purpose of this study was to examine whether standard clinical outcome measures that have been validated in older adults to identify falls risk would adequately identify falls risk in individuals with visual impairment due to RP. This was accomplished by examining the relationships between falls history and clinical outcome measures.

Methods

Participants

Community-dwelling adults from metropolitan Atlanta and the surrounding area were identified via chart review at the Atlanta Veterans Affairs Medical Center (VAMC) Eye Clinic and Emory University Department of Ophthalmology and then sent letters of recruitment. Potential participants were also recruited via flyers and presentations at local organizations providing services to individuals who are blind or visually impaired, including the Center for the Visually Impaired in Atlanta, and the Georgia chapter of the Foundation Fighting Blindness. Inclusion criteria for the study were: an established diagnosis of RP or Usher's syndrome, adult age (between 18 and 90 years), legal blindness according to visual field standards (i.e., central visual fields less than 20 degrees in diameter, with nearly complete to complete peripheral visual field loss in the better eye), ability to walk household distance without an assistive device, and ability to stand for at least 20 minutes. Individuals were excluded if they had binocular visual acuity worse than 20/800, more than mild (1+) cataracts, other eye diseases (including glaucoma or diabetic retinopathy), history of eye injury, insulin-dependent diabetes or serious nonophthalmic diseases (e.g., Parkinson's disease) that might affect participation in the study. This study adhered to the tenets of the Declaration of Helsinki, and was approved by the institutional review board of Emory University and the Atlanta VAMC Research and Development Committee. Informed consent was obtained from all participants after the nature and possible consequences of the study had been explained.

Study Protocol

This study was part of a larger longitudinal study investigating everyday visual function in people with RP. The protocol involved two days of extensive

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assessment of visual impairment, functional vision, and gait and balance, in addition to self-report measures of balance-related confidence and visual abilities. The protocol included tests of visual impairment such as near visual acuity, low luminance acuity, and color vision as well as functional vision tests, such as face recognition, visual search, and reading. The results presented here concentrate on distance visual acuity, contrast sensitivity, and visual fields, along with balance and gait testing. Participants were compensated for their time and travel expenses.

Materials and Procedure

Vision Test Battery

The measurement of visual acuity and contrast sensitivity was performed with binocular viewing. Participants wore their habitual distance refraction. Visual acuity (logMAR) was measured with the Early Treatment in Diabetic Retinopathy Study (ETDRS) chart (Ferris, Kassof, Bresnick, & Bailey, 1982) at 3 meters or at 1 meter if the participant was unable to read all of the letters on the top two lines at 3 meters. Letter-by-letter scoring was used, as recommended for best test-retest reliability (Bailey, Bullimore, Raasch, & Taylor, 1991), and testing continued until the subject gave five consecutive wrong answers. Contrast sensitivity values were measured with the Pelli-Robson chart (Pelli, Robson, & Wilkins, 1988) at 1 meter, also with letter-by-letter scoring (Elliott, Bullimore, & Bailey, 1991). Visual fields were measured with the Humphrey Field Analyzer using the 81-point full-field 3-zone screening protocol. Eight of the 10 participants also had macular perimetry performed with the scanning laser ophthalmoscope (SLO) to determine the extent of the central visual field (Sunness, Schuchard, Shen, Rubin, Dagnelie, & Haselwood, 1995).

Balance and Gait Test Battery

Gait speed was assessed by having participants walk at their preferred and fast gait speed over a level indoor surface without an assistive device. The time to walk 6 meters was recorded using a stopwatch (Bohannon, 1997). The participant began the trial 1.5 meters behind the start point for the 6-meter distance and continued walking for 1.5 meters past the end of the 6-meter distance. Timing began when the first foot crossed the start point and ended

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when both feet crossed the end point. Participants were instructed first to walk at their "normal preferred pace" and then they repeated the task with the instructions "to walk as quickly as safely possible." Participants wore a safety belt and were accompanied by either a physical therapist or trained research assistant to ensure safety. Three trials of each instructed speed were performed and average speed was calculated.

The ability to adapt gait in the presence of external demands was assessed using the Dynamic Gait Index (DGI). The eight items of the DGI include walking while changing speed and turning the head, walking over and around obstacles, and stair climbing (Shumway-Cook & Woollacott, 2001). Scoring of the DGI items is based on a 4-point scale from 0 to 3, with 0 indicating severe impairment and 3 indicating normal ability. A maximum total score of 24 is possible and scores less than 20 indicate high risk for falling (Shumway-Cook et al., 1997; Whitney et al., 2000). The DGI has excellent inter-rater as well as test-retest reliability (0.96–0.98; Shumway-Cook et al., 1997).

The Sensory Organization Test (SOT) was used to assess the integration of sensory information for balance by measuring postural sway under conditions in which visual and somatosensory feedback is altered (Nashner, Black, & Wall, 1982). The SOT is organized into a series of six conditions of increasing difficulty. The first three conditions are performed on a firm surface with eyes open (condition 1), eyes closed (condition 2), and finally with vision sway-referenced (condition 3). The final three conditions are performed with the support surface sway-referenced with eyes open (condition 4), eyes closed (condition 5), and with vision sway-referenced (condition 6). Sway-referencing refers to either the visual surround or the support surface moving in the same direction and amplitude as the person's sway. Sway-referencing provides inaccurate visual or somatosensory input. The SOT composite score has been found to have good test-retest reliability, and to differentiate between fallers and nonfallers in community-dwelling older adults (Ford-Smith, Wyman, Elswick, Fernandez, & Newton, 1995).

Self-Report Measures

Falls history was based on self-report. Participants were asked to respond to the question "How many times have you fallen within the past year?"

Participants were asked the cause of each fall to determine the mechanism of the falls where possible.

The Veterans Affairs 28 questionnaire (VA-28; De l'Aune, Welsh, & Williams, 2000) focuses on vision-related everyday function, or activities of daily living (ADLs). This self-reported assessment of ADLs was chosen because it was developed and validated specifically for individuals who are legally blind. The questions deal with activities that are important and can still be performed by those with severe vision loss or worse, such as outdoor chores, paying bills, or using public transportation. In addition to rating the difficulty of each activity, subjects were asked to rate the importance of the activity, their sense of independence for performing the task, and their satisfaction with their ability to perform the task. The self-report questionnaire was verbally administered by the research assistant. Of the 28 questions, the analysis for this study concentrated on the answers to the orientation and mobility (O&M) subscale items (11 questions, including how difficult is it for you to "generally avoid obstacles as you walk?"). The answer choices were: 1 through 4 (not difficult at all to impossible) or U (do not perform task due to non-visual reasons). Answers of U were counted as missing data. An overall average difficulty score for all the items of the VA-28 scale was calculated as well as an average difficulty score for the O&M subscale.

The balance self-efficacy scale (BES) is a scale that measures an individual's current balance-related confidence while performing a variety of ADLs. These include getting in or out of a bed, chair, or shower; walking on uneven surfaces (with and without good lighting, with and without assistance); and going up and down stairs with or without a railing (Gunter, De Costa, White, Hooker, Hayes, & Snow, 2003). Subjects rated their confidence in their ability to perform different activities without losing balance. There are 18 items in the BES, and each is rated on a scale from 0 to 100 percent (0 indicates no confidence, 100 indicates absolute confidence). Average overall confidence across the 18 items was calculated. BES scores have been correlated with balance and mobility as well as faller status in older women (Gunter et al., 2003).

Data Analysis

Descriptive statistics were used to summarize the characteristics of the sample. Bivariate correlations (Pearson's) were performed to examine the relation-

Table 1. Participant Characteristics (n = 10)^a

	Median	Range
Age (years)	51.0	31–80
Gender	6 females; 4 males	
Number of falls in past year	1	0–3
Visual acuity (logMAR)	0.47	1.32–0.00
Contrast sensitivity (logContrast)	1.00	0.10–1.65
Visual field diameter (degrees)	8	2–18
VA-28 total score	1.80	1.45–2.93

^a VA-28 = Veterans Affairs 28 questionnaire.

ships between number of falls (by self-report history) and clinical measures of visual function and balance and gait. Due to the exploratory nature of the study, variables that accounted for at least 16 percent of the variance ($r > 0.4$, the strength of which is considered moderate) were considered significant. Data were analyzed using PASW Statistics (formerly SPSS), version 18.0.

Results

Participant Characteristics

Ten participants with established RP completed tests of vision and gait and balance (Table 1). One participant had been diagnosed with Usher syndrome type II, which causes congenital hearing loss, but spares vestibular function, unlike type I which results in vestibular deficits (Yan & Liu, 2010). That participant had rotary chair testing to confirm normal vestibular function. One participant was missing contrast sensitivity and visual field scores and one participant was missing BES score. The median age of the participants was 51.0 years. Given the small sample and broad age range, the actual ages of the participants are listed here for reference: 31, 36, 39, 49, 50, 52, 53, 59, 74, and 80 years. Six of the 10 subjects were female. The monocular visual fields were very similar between eyes for each participant. Since these subjects use the fovea, it is reasonable to overlap the monocular visual fields to determine the extent of the binocular visual field. The Humphrey visual fields confirmed that each of the participants had nearly complete to complete peripheral visual field loss and less than 20-degree central visual fields. The participants showed a range of visual acuity and contrast sensitivity values from age-normal to abnormally low.

Of the 10 participants, eight participants had experienced at least one fall (range: 1–3) for a total of 13 falls within the previous year. No reason was given for the majority of falls ($n = 8$ falls); otherwise, tripping was the only reason listed as causing the fall ($n = 5$ falls). Participants were asked to complete information on a health questionnaire regarding assistive device use. One participant did not use any assistive device, eight participants used a white cane (as a visual guide, not as a support cane), and data are missing for one participant. See Table 1 for complete demographic information.

Relationship Between Clinical Measures and Falls History

In spite of the majority of participants having experienced a fall in the previous year, the clinical assessments of balance and gait were normal for the majority of participants (Table 2). Six of the 10

Table 2. Balance and Gait Abilities of Participants^a

	Median	Range
Preferred gait speed (meters/second)	1.10	0.87–1.49
Fast gait speed (meters/second)	1.68	1.16–2.08
Dynamic gait index (out of 24)	20	13–22
Sensory Organization Test (out of 100)	71	53–86
VA-28 O&M subscale	2.05	1.45–3.09
BES (percent)	78	65–97

^a VA-28 = Veterans Affairs 28 questionnaire; O&M = orientation and mobility; BES = balance self-efficacy scale.

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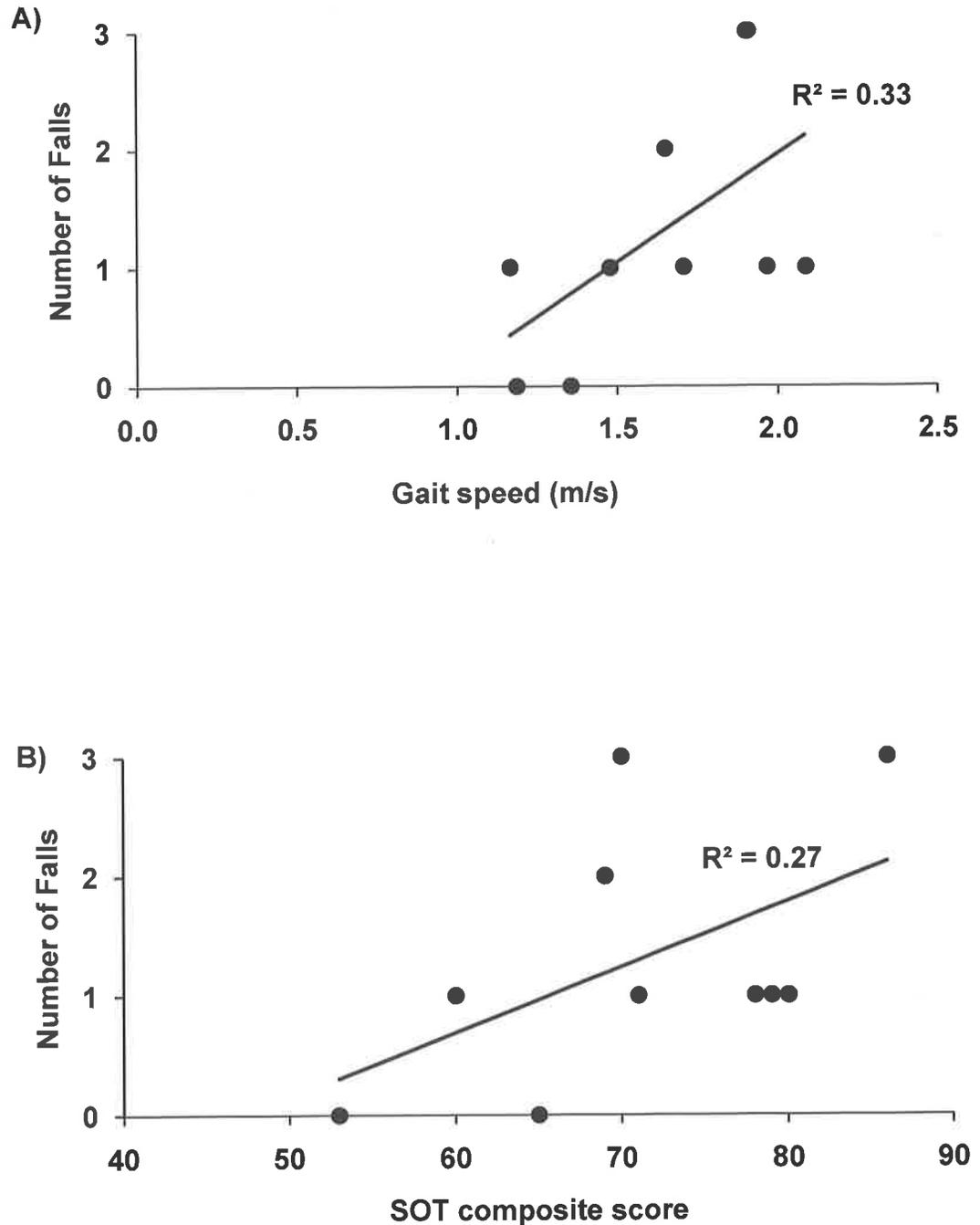


Fig. 1. Relationship of falls history (i.e., the self-reported number of falls per person for the past year), with respect to the distribution of the variable for individual participants. A) fast gait speed (meters/second); B) Sensory Organization Test (SOT) composite scores; C) Dynamic Gait Index (DGI) scores; D) visual field diameters (degrees). The line for linear regression and the value of R^2 are displayed for each variable.

participants had normal preferred gait speed, and 8 of 10 had normal fast gait speed based on age and gender reference values (Bohannon, 1997). None of the participants met the criteria for falls risk based on gait speed (< 0.70 meters/second; Monterro-Odessa

et al., 2005). Seven participants had normal SOT scores for age (NeuroCom International, 2001). Six of the participants were classified as low risk for falls and four as high risk for falls based on their total DGI score.

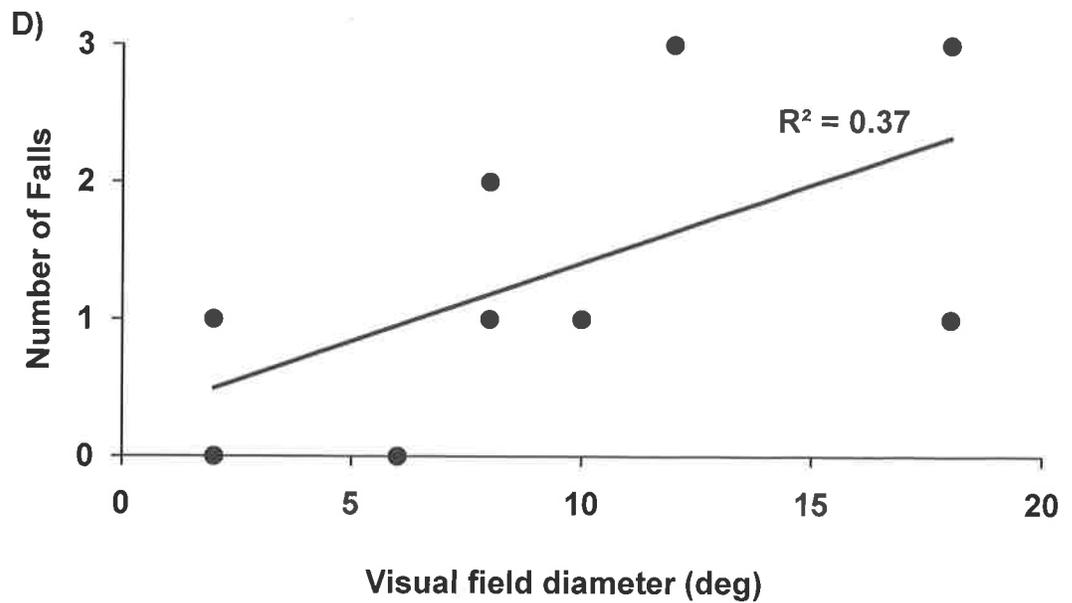
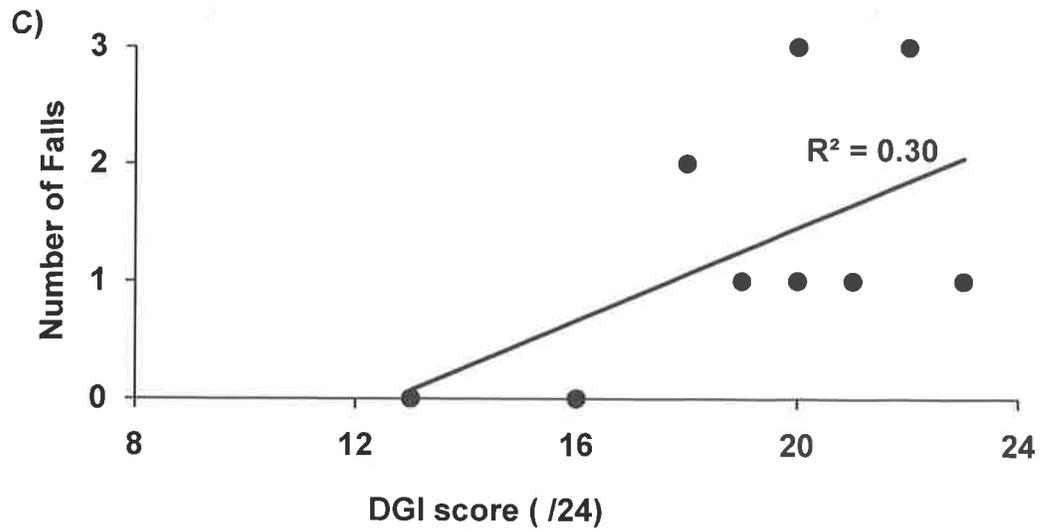


Fig. 1. Continued.

The number of falls in the previous year was moderately correlated to fast gait speed ($r = 0.57$, $p = 0.084$; Figure 1A), SOT composite score ($r = 0.52$, $p = 0.01$; Figure 1B), DGI ($r = 0.55$, $p = 0.010$; Figure 1C), and visual fields ($r = 0.60$, $p = 0.08$; Figure 1D). The positive direction of these relationships (Figure 1A–D) indicates that a greater

number of falls was related to faster gait speed (generally indicative of better mobility), higher SOT composite scores (higher scores indicate greater stability), higher DGI scores (higher scores indicate greater dynamic stability and lower falls risk), and larger visual fields. It was expected that the correlations between falls history and these variables

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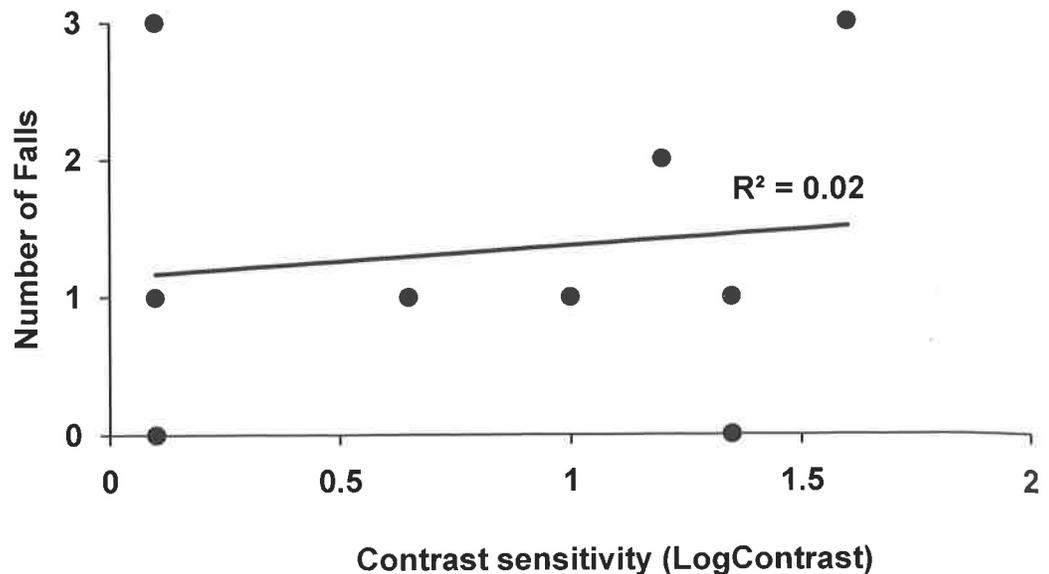


Fig. 2. Relationship of falls history with contrast sensitivity. The line for linear regression and the value of R^2 are displayed.

would be negative; that is, a greater number of falls was expected to correlate to slower gait speed (indicative of worse mobility), lower SOT (indicative of greater instability), lower DGI (indicative of worse dynamic stability) scores, and smaller visual fields (indicative of greater limitations of peripheral vision). All other correlations with the number of falls (including age, BES, VA-28, and O&M subscale scores, preferred gait speed, visual acuity and contrast sensitivity) were small ($r < 0.40$; e.g., Figure 2) and thus considered not significant.

Discussion

In this group of adults with RP, the incidence of falls over the previous year was 80 percent. It is well recognized that falls are a problem in children and older adults, but not typically for adults between those two age groups, such as half of our participants who were in the middle-aged group. It is estimated that one-third of adults aged 65 and older will fall in a given year and that number increases with increasing age (Hausdorff, Rios, & Edelberg, 2001). Thus, the finding that 80 percent of our sample fell in the previous year was much higher than would be expected based on findings from normally sighted adults. Turano et al. (1999) also reported a high rate of falls among their survey respondents (46 percent), although not as high as in our study, and Black et al. (1997) cited tripping and falling as "commonly

reported mobility problems" for individuals with RP. In combination with the present results, the findings from these studies suggest that much work remains to identify whether there are specific impairments that contribute to falls and how interventions might be tailored to individuals with RP in order to reduce falls incidence. Further studies should clarify the risk factors specifically for injurious falls in people with visual impairment in order to prevent injuries.

In the present study, the falls in which a mechanism could be determined were related to trips. Studies that have examined the types of falls that occur in older adults have identified the base-of-support disturbance (i.e., slips or trips) to be the most common cause of a fall, with trips being associated with approximately 40 percent of falls (Freeman et al., 2007; Maki, Holliday, & Topper, 1994; Nachreiner, Findorff, Wyman, & McCarthy, 2007). Based on the O&M literature, it was not unexpected that tripping would be a primary cause of falls in individuals with RP since reduced visual fields and/or reduced contrast sensitivity may decrease the observance of obstacles (e.g., Black et al., 1997). In addition, impaired night vision in RP could also increase falls incidence, again due to not seeing obstacles in dim lighting, both indoor and outdoor (Hartong et al., 2004).

Falls in older adults are known to be caused by multiple factors, typically resulting from the interaction of risk factors, which makes falls prediction a

complex task. In this study, each potential factor was explored independently (e.g., visual acuity or gait speed) when it is likely that more than one factor contributed to a fall. In addition, the specific factor(s) contributing to falls may be different for different people. Still, it was surprising that results from standard clinical tools used to identify falls risk in older adults and adults with impaired balance either did not correlate with falls history in this sample, or correlated in the opposite direction to expectations. For example, slower gait speed has been related to greater instability and increased falls incidence in older adults (Monterro-Odasso et al., 2005). However, in the present sample, the two individuals who had not fallen in the previous year had the slowest gait speed for the fast condition. Likewise, lower SOT and DGI scores were associated with fewer falls. It may be that some individuals with RP adopt a more conservative gait pattern and by slowing down, they allow more time to preview the environment and therefore avoid obstacles in the path. This is a common mobility strategy taught by O&M instructors (Geruschat & Turano, 2002). Risky behavior (fast walking speed) may predispose individuals to falls following a trip. Older adults with faster gait speed who are tripped are more likely to fall (Pavol, Owings, Foley, & Grabiner, 2001). It has been demonstrated in the literature that, as a group, people with RP walk more slowly than normally sighted peers (Black et al., 1997; Geruschat et al., 1998; Turano et al., 1998). However, in the present study, we found that 6 out of 10 participants walked within normal limits for age and gender at preferred speed, and 8 out of 10 at fast speed (Bohannon, 1997). Differences between the findings from the current study and these others may in part be attributed to a lack of matching on gender and differences in walking routes. For example, in the Black et al. study (1997), the control group had twice as many males as the group with RP, and females on average have a slower gait speed. Geruschat et al. (1998) timed walking on a route that involved avoiding overhead obstacles to compare walking speeds between groups, whereas the current study used a straight path without any obstacles to calculate gait speed and then compared those values to reference values in the literature. The findings in this study, while preliminary, serve as a reminder that criteria for falls risk based on outcome

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measures (such as gait speed, DGI, and SOT) need to be validated for specific groups, including those with visual impairment resulting in severely reduced visual fields. The majority of work in the area of falls prevention has been limited to older adults, rather than younger adults with specific disabilities.

Limitations

The sample of this study was small, so generalizations should be made with caution. In order to develop appropriate interventions to prevent falls in individuals with visual impairment, we have to understand the relationship between various types of visual impairment (e.g., central vs. peripheral visual loss), and balance and gait as it relates to falls risk. In this manner, appropriate interventions can be targeted to the specific balance and gait impairments. Alternatively, for some types of visual impairment (e.g., RP) it may be more critical to have appropriate O&M training in order to compensate for the loss of visual fields and to prevent falls, rather than targeting sensorimotor deficits, such as strength issues or sensory integration issues.

Future Directions

In order to identify the most appropriate approach to preventing falls in individuals with visual impairment, prospective studies should be done. These studies would aim to identify patterns of falls (i.e., trips, slips, or other), particularly with respect to injurious falls, by using falls diaries and assessing multiple measures of physical performance and more specific self-report measures of mobility (e.g., Turano et al., 1999). The outcomes of such research would inform the practice of physical therapists as well as O&M specialists by identifying specific areas of deficit and developing potential strategies (e.g., walking more slowly) to avoid future falls based on the deficits identified. Gathering additional information regarding past O&M training would also be useful, in order to determine its impact on falls incidence in people with RP. The question remains as to which assessments will be most helpful and whether new performance criteria need to be developed for people with visual impairment. While there was a significant relationship between visual fields and falls history in the present study, the relationship was in an unexpected direction. Furthermore, there was no significant relationship between falls history and contrast sensitivity for this (small)

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group. Based on previous literature, these findings were unexpected and deserve further study to clarify the impact of age and other factors on the results.

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