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Disorientation and loss of wayfinding in individuals with congenital blindness and other affecting comorbidities

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Abstract

Orientation and navigation skills are considered as crucial prerequisites for efficient wayfinding in the surrounding environment and in community both for sighted and blind individuals of all ages. In case of partial or total loss of vision, navigation and wayfinding become more challenging. The acquisition of proficient orientation and navigation by visually deprived individuals is possible when accurate guidance and training are available, and the central nervous system is intact. In cases of head trauma, neurological lesions (specifically, right hemispheric lesions), or genetic factors, the acquisition of orientation and navigation becomes a disturbed process. In some severe cases, the individuals become, literally, "space-less." This profound disorientation leads to inefficient functional outcomes, especially when the environment is unfamiliar. These spatially disoriented individuals with loss of independence in both indoor and outdoor wayfinding skills need a long-term, persistent individual guidance by caregivers to prevent risks, a reality that limits daily functions and participation in life roles. Alternative and differential strategies for individual training methods of teaching orientation and navigation are needed. This article discusses this issue on the basis of the theoretical level and proposes practical methods and strategies for enhancing the very initial foundations of orientation and navigation for this specific "space-less" group. Functional impacts and practical implications are further discussed.

Keywords

Differential strategies, navigation skills, spatial orientation, wayfinding

Introduction

Wayfinding is a function that enhances human occupations, promotes participation in community activities, and enables life roles in both indoor and outdoor environments. This function is crucial for sighted people and for those who have severe visual impairment or total blindness (Fortin et al., 2008).

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Individuals with severe visual impairment or total blindness face daily challenges during the performance of routine tasks and participating in various life roles (Giudice & Legge, 2008). Indoor and outdoor mobility and navigation, in both familiar and unfamiliar environments, need multimodal processing skills (e.g., auditory, tactile, vestibular) and motor praxis abilities that are supported by cognitive competencies and executive functions (e.g., selection of the navigation strategies and performance of the motor sequence) to compete with dynamic, everchanging environmental changes (Schinazi, Thrash, & Chebat, 2016).

The concept of Wayfinding, as described by Montello (2005), may describe, in a summarized manner, the whole spectrum of complex, multimodal task of locomotion, orientation, and navigation. Wayfinding integrates the sensory, motor, and cognitive skills with the executive functions that are necessary for proficient performance of transferring from location A to location B in any environment. Wayfinding involves ongoing reasoning process regarding immediate and remote environments and requests the application of transient short-term and long-term mental representations. During Wayfinding, people may also adopt reference frames other than those directly experienced. In contrast, the concepts of "locomotion" and "mobility" relate to the immediate responses to environmental features, such as avoiding a bench in the middle of the route or avoiding stepping over an edge. These immediate responses are primarily egocentric because environmental information is acquired with reference to the observer's body (Schinazi, Thrash, & Chebat, 2016). In wayfinding, the individual must be aware of two main types of reference frames during locomotion and mobility: egocentric and allocentric reference frames. Allocentric reference frame always involves at least one individual, two environmental features, and the spatial relations among them, while egocentric reference frame involves one individual, one concrete environmental feature, and the spatial relation between both.

Spatial navigation and wayfinding

Indoor and outdoor navigation under the condition of deprivation of functional vision, due to congenital conditions or acquired vision loss, demands continuous decision-making skills, executive functions, attention to immediate stimuli, and reevaluation of ongoing locomotion from an initial location to a defined destination. Individuals with blindness confront navigation difficulties and orientation problems that need complex and abstract problem-solving skills, and thus they are forced to adapt to their environment by finding alternative resources and effective strategies for route learning (Fortin et al., 2008). These individuals with vision loss rely on other immediate sensory input to execute their adaptive motor responses toward near-surrounding features (i.e., one step on a stair up or down the hallway at home) or to consider features of the more distal parts of the environment (i.e., approaching toward cross-roads with traffic lights). In both environmental conditions, wayfinding needs continuous orientation and attention to the everchanging multimodal sensory information that is processed dynamically for the purpose of execution of motor actions and adaptive behavioral responses (Giudice & Legge, 2008).

Previous studies that have investigated both navigation strategies and the effectiveness of navigation performance have typically constrained blind and sighted participants to adopt similar strategies. For example, this occurs when sighted participants are asked to wear blindfolds or when blind participants are asked to complete visually guided tasks (e.g., pointing judgments; Haber, Haber, Pennigroth, Novak, & Radgowski, 1993). In these cases, and similar research procedures, a difference in performance is inconclusive with respect to abilities alone because the strategy that was adopted by both blind and sighted people invariably disadvantages one of the groups. These results are basically inconclusive because researchers are unable to detect a difference in performance.

Therefore, study procedures should apply differential strategies to either groups of participants congruent to vision status (i.e., sighted or blind) to gain conclusive results.

Some studies investigated the hypothesis whether individuals with blindness have an advantage over sighted, or even blind-folded sighted, individuals. Fortin et al. (2008) found that individuals with congenital blindness ($N=12$; M age = 33.8 years) or even individuals with late onset of blindness ($N=7$; M age = 39.9 years) significantly have superior navigational skills on a route-learning task and a pointing task in a maze compared to sighted, matched controls ($N=19$). Moreover, the researcher proved that the Hippocampus of those participants with vision loss have developed a larger volume in its posterior and anterior parts, indicating a remarkable plasticity in this specific brain structure that is well known to be involved in spatial procession.

As reported by numerous individuals with blindness, any occurring of inattention or transient distraction may lead to an immediate decrease in the effectiveness and accuracy of wayfinding. In some cases, individuals with blindness have additional neurological disorders and cognitive dysfunctions that may affect their navigation and wayfinding skill acquisition, such as children who had shunt surgery or recurrent surgeries for shunt revisions (e.g., Jarjoura, 2018) and children with genetic syndromes influencing the central nervous system (e.g., Hippocampus; Fortin et al., 2008). This population confronts dual conditions: the total loss of vision and the total loss of navigation, and wayfinding abilities which lead to relying on caregiver assistance. These individuals, although considered as a minority among the population of people with visual impairments, have numerous functional problems in their daily routine, both in the indoor (i.e., dressing, locomotion, Braille printing and tactile discrimination, etc.) and in the outdoor tasks (i.e., community mobility, wayfinding in unfamiliar areas, etc.) (Jarjoura, 2018). Such foundational dysfunctions, especially in young children who did not acquire the minimal residual representations of their immediate surrounding space of living, propose the concept of "Space-less" children which refers to the absence of mental spatial-perceptual representations even for daily, familiar locations and destinations. This situation of absent spatial frame-of-reference for the young child may provide an explanation for the common repetitive behavior of spinning around self and rocking back-and-forth that characterizes some young children with congenital blindness and additional diagnosis and comorbidities. In other words, these children seem to lack any frame-of-reference for their locomotion in the immediate environment, although familiar and explicit; thus, they tend to respond "on-spot" due to an absence of any minimal directional information or any concrete route features. Although the use of a Cane was proved to be effective and enhances mobility skills and navigation (Serino, Bassolino, & Farne, 2007), children who lack any spatial reference still confront foundational difficulties in navigation and wayfinding even when the Cane is used manually. In other words, no computational processing is available for the child's spatial cortices to compute. As a result, spinning-around-self seems to become the "default" situation available in this status. In the absence of mental representations of short routes and conceptual directions of the immediate concrete spaces, spinning-around-self becomes the only possible independent "mobility" function.

Some previous studies reported that repetitive movements, such as body rotating and rocking, are believed to be evoked by sensory modulation disorders (e.g., sensory sensitivity) that are common in autism and other conditions (Brown, Hobson, Lee, & Stevenson, 2006; Hsu & Ho, 2009), especially those related to low IQ and more severe cases (Bishop, Richler, & Lord, 2006; Militerni, Bravaccio, Falco, Fico, & Palermo, 2002). Such repetitive body movements are usually explained as self-stimulatory behaviors and statistically correlated with anxiety and psychological stress (Rodgers, Glod, Connolly, & McConachie, 2012; Wigham, Rodgers, South, McConachie, & Freestom, 2015).

Fazzi et al. (1999) mentioned that prolonged hospitalization, motor limitations, reduced capacity for exploration, and reduced sensory stimulation are common factors that are believed to evoke repetitive and stereotyped behaviors by congenitally blind children. When considering the correlation between anxiety, stress, and repetitive behaviors in congenitally blind children, clinical observations and other observations in various natural life environments reveal no indications for external factors of anxiety nor psychological stress. Internal factors may exist, although such factors are not believed to play significant roles in evoking repetitive behaviors. The study of Rogers and Newhart-Larson (1989) revealed that blind children due to Leber's congenital amaurosis showed significantly higher frequency and more severity of autistic behaviors compared to congenitally blind children due to other causes and conditions according to Childhood Autism Rating Scale (CARS) and the Autism Behavior Checklist (ABC). Such findings suggest hereditary genetic factors rather than psychological factors.

Alternatively, other factors and interpretations may be possible in the case of children with congenital blindness, especially those who are young and still did not acquire basic mobility and orientation skills. One may hypothesize that this specific group of children merely have their own body position-in-space as their only egocentric frame-of-reference for mobility and motor behavior. Those "space-less" children can be compared to the condition of astronauts that are navigating in a dark space and, thus, lack any minimal knowledge regarding orientation and navigation from initial location to a specific destination in such space. Consequently, wayfinding is impossible. In such space-less conditions, the "default" situation becomes only performing "on-spot" movements through rocking, rotation of upper trunk, and spinning around self.

Based on the above description and interpretation, alternative strategies are needed to be developed and specific implementation of such novel strategies must be evaluated and modified to enable these individuals to function at their homes and at local community. This article tries to propose a variety of novel orientation and navigation strategies that are believed to improve wayfinding competencies to a functional level. The strategies are differential and adapted to individual differences in spatial orientation status and limitations in wayfinding, of those who have partial orientational skills and those who lack minimal mental representations of the surrounding environments (space-less individuals).

Assessment of spatial wayfinding components

The development of adapted strategies for navigation and wayfinding is based on a two-step process: the first step is to determine whether the individual with blindness and spatial disorientation totally lack any mental representation of their daily spaces (space-less) or they gained an inefficient performance of locomotion and wayfinding in a familiar environment. To refer an individual to a specific group, a comprehensive evaluation of the spatial-navigational and wayfinding skills should be administered using observations in various living environments during spontaneous and guided locomotion, interview with the parents, and navigational tasks based on verbal (i.e., auditory) and haptic (i.e., tactile-motor) information.

Observations

During observations in various environments (home, kindergarten, school, play yard, etc.), the observer should take notes regarding the following behaviors: (1) the existence of spinning around self and/or rocking self back-and-forth, (2) initiation of movement toward a nearby purposeful destination (i.e., entrance, bed, sink, etc.), (3) staying in a very limited space while seated or sitting on ground for long period of time, (4) turning head and upper trunk toward a source of auditory stimulation behind the child, and (5) correcting self-locomotion route when walking into an obstacle in the middle of the route (i.e., furniture, wall, persons, etc.).

Interview with the parents

Understanding the evolving foundations of navigation and wayfinding skills needs specific information that only the parents, or the primary caregivers, can deliver explicitly through a semi-structured interview targeting some specific aspects of the child's behaviors and competencies at home. In some circumstances, the children spend more time with their primary caregivers than with their own parents and those caregivers become more familiar with the children's behaviors, routines, and characteristics and even deeply influenced by the caring burden and stress to the level of moderate to severe depression (Dada et al., 2013).

The questions to the primary caregivers may focus, mainly but not only, on the following domestic functions and behaviors: "Does the child get off the bed and initiate spontaneous locomotion inside the room?"; "Does the child find the bedroom's door independently?"; "Does the child climb on bed independently and then locate and put the head on the pillow?"; "Does the child call mother's/father's name and move around in searching behavior?"; "Does the child reach out with arms toward the parent's location forward or to one side?"; "Does the child approach toward the house's entrance door when open and sun shine illuminate the entrance or when the wind blows through it?" When entering home, "Does the child head directly toward a specific destination?"; "Does the child seem to recognize the toilet's space and concrete features?"; "Does the child listen to TV while face heading toward it, or in an angle?"; "Does the child open and close a drawer independently while locating an item inside it?"; "Does the child understand and use the concepts 'right-side' and 'left-side' correctly?"; "Does the child understand and use the phrases '. . . in front of you' and '. . . on your back' correctly?"

These questions, and other similar and relevant ones, help the professional to create a wider picture regarding the child's spatial orientation, navigation, and wayfinding skills. Integrating the interview's outcomes with the observations' outcomes is expected to illuminate the "spatial status" of the child.

Spatial navigation and wayfinding tasks

The following proposed tasks target gathering information regarding the initial foundations of navigation and wayfinding. The tasks are explicit and can be administered by any professional that have a minimal work experience with individuals with blindness: (1) The child approaches toward the caregiver located 2 m ahead, who is calling the child's private name repetitively; (2) the child initiates locomotion between two persons, back-and-forth, with a maximum of 1 m distance between both; (3) the child navigates between a familiar location A and nearby familiar location B, about 1 m apart, back-and-forth, independently (i.e., between chair at classroom and a drawer containing toys); (4) when located next to a door, the child locates and opens the door's knob and walks out; (5) the child walks around an obstacle in the middle of the navigation route and continues walking; (6) the child keeps the Cane on the ground consistently without lifting and waving it frequently in a random manner; (7) when asked about the location of items in relation to self, the child's answers include the congruent spatial concepts: "in front of me," "on my back," "on my right side," "on my left side."

Differential strategies for spatial wayfinding performance

Based on the previous assessment outcome measures and on evidence-based data, an individual intervention plan and differential strategies should be prepared and adapted to the level of spatial orientation, navigation, and wayfinding that was determined by the assessments' results: a disturbance of spatial competencies versus a total absence of such competencies. In each case, different strategies are proposed.

Absence of spatial competencies

Children who are observed to be totally deprived from spatial-perceptual and navigational skills, and evidenced to be lost in their immediate environment, seem to lack any spatial frame-of-reference for their locomotion in the personal, egocentric motor-haptic level. Such children need to acquire the very initial foundations of spatial-perceptual orientation and first steps of navigation starting from egocentric frame-of-reference. The caregiver plans and performs various tasks that are based on the principle of one child–one environmental feature structure. The environmental feature could be the caregiver of a concrete item within hand reach. Following are some practical tasks that are designed on the basis of this principle. These tasks are sequential and developmental, that is, arranged from simple to more complex, and from constricted, egocentric space to a wider space in a familiar environment. As the caregiver instructs the child in each of the following tasks, no additional guidance or clues are allowed for the child to uncover the real status of the spatial foundations.

1. Asking the child to locate and reach out hands toward the caregiver, while the latter is changing his or her location around the child, and then the child is asked to name the direction: "In front of me," "at my side," "at my back," "at my right side," "at my left side." The caregiver can use auditory stimulus (i.e., a tiny bell, tongue clicks, finger clicks, etc.) to enhance the child's response.
2. The child is seated in the middle of four desks around him, where on the top of each desk, there is a familiar item that was previously recognized and named by the child. Then, the child is asked to locate each item according to naming it by the caregiver. Later, the caregiver instructs the child to determine the location of the specific item as named randomly by the caregiver. The child is asked to name the direction: "in front of me," "at my side," "at my back," "at my right side," "at my left side."
3. The caregiver accompanies the child along the short route between the seat and the door, back-and-forth. And then, the caregiver instructs the child to get up from the seat and to approach toward the door and get back to the chair again.
4. The caregiver posits the child on the door's opening and instructs the child "to turn right," or "to turn left," and continue walking at that direction. The task should be repeated six times, at least, to assure that the child has understood and performed the task correctly and accurately.
5. The caregiver posits the child on the door's opening and instructs the child to walk few steps straightforward, and then asked to turn back to the door's opening, independently.
6. The caregiver posits the child in front of the stairway from ground floor to the first floor and instructs the child to start walk up the stairs while supported by the handrails or by the wall.
7. The caregiver posits the child in front of the stairway from the first floor to the ground floor and instructs the child to start walk down the stairs while supported by the handrails or by the wall.
8. The caregiver posits the child in the middle of the hallway in a familiar floor and then instructs the child to choose a preferred location in a specific direction to walk toward it. Then, the caregiver instructs the child to start walking toward that location by himself.

Non-proficient spatial competencies' navigation skills

In some cases, children may have functional orientation skills and distorted navigational competencies that lack accuracy and speed. In these cases, due to medical condition, genetic factors, or neurological disorders (i.e., shunt surgery in the right, spatial hemisphere; Jarjoura, 2018), blind children lack foundational spatial orientation and navigation skills even in a familiar environment in which they experience their routines repetitively. This group of children with blindness needs complementary activities and tasks that, mainly, enhance accuracy and speed. The following proposed activities represent a wide spectrum of skills and competencies that target both accuracy and speed of performance in navigation and wayfinding functions. For organizational reasons, these activities are presented in an integrated manner between accuracy and speed because both criteria cannot be practically separated during activity performance, as follows:

1. The caregiver instructs the child to get up from classroom seat and head toward the door, open the door's knob and close it again, and then get back to seat within countdown from 10 to 0 by the caregiver.
2. The caregiver instructs the child to get up from classroom seat and walk toward the teacher's seat, take a Braille paper, and get back to seat within countdown from 20 to 0.
3. The caregiver instructs the child to get up from seat, head to classroom's door, open it, and turn right. Then, walk 10 steps and turn back all the way to seat, within count range from 1 to 30.
4. The caregiver instructs the child to walk along the route from classroom seat to the toilet entrance of the same floor, and then turn back to classroom seat within count range from 1 to 60.
5. The caregiver instructs the child to walk along the route from classroom seat to the principal's office, independently, within a limited period of time that is relevant to each school building.

Summary

This article aimed to focus on the spatial orientation and navigation difficulties and, in specific cases, total absence of such skills that significantly limit the wayfinding competencies of some children with congenital blindness (space-less children). These children usually have other genetic conditions or neurological disorders that directly limit their spatial foundations compared to the baseline level. Repetitive behaviors, such as rocking own body and spinning around self, are not believed to be comparable to those observed in children with autism spectrum disorder, due to profoundly different factors such as IQ level (Brown et al., 2006) and genetic conditions (Rogers & Newhart-Larson, 1989). With the lack of such navigational skills, these children are restricted to a very limited space and, thus, their participation in daily functions is inefficient and, needs a continuous, long-term caregiver assistance. To decrease this burden on the caregivers, differential strategies should be applied individually to help these children to acquire satisfactory functional navigational skills and wayfinding competencies. To achieve this aim, accuracy and speed of these skills must be improved. Applying performance time limits and demanding higher accuracy of performance from this target population may enhance their performance and help them acquire a significantly more proficient wayfinding skills in their familiar environment. The burden on caregivers is expected to decrease significantly, while the children's level of participation in adapted activities and life roles are expected to increase.

Future studies should investigate the effectiveness of a wide spectrum of differential navigation and wayfinding strategies in congenitally blind children especially for those who are "space-less" and have additional genetic conditions (i.e., Leber's congenital amaurosis). Similarities in repetitive motor actions and stereotyped behaviors between this group of children and children with autism spectrum disorder should be deeply and comprehensively investigated to achieve new evidence for the existence of significantly different factors for such behaviors between both groups.

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