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Journal of Clinical Research and Ophthalmology Pertechz



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Introduction

Typically when someone loses a sense due to trauma, injury or disease, the loss is considered to be deprivation and the person is often labelled with a disability. Loss of a body part not only impacts functionality but it can also affect the individual cosmetically and, in turn, affects the individual's psychological and psychosocial wellbeing. Historically, restorative measures towards the sensory loss has focused on the perception that it is a loss, rather than a change that may, in fact, leave the person with a different set of senses as compared with fewer senses. This has been because our understanding of the environment has placed great emphasis on the reliance of sensory feedback [1].

However, there is a growing trend towards regarding sensory losses as changes to lifestyle that render the person with a new growth and direction for life goals, termed post-traumatic growth [2], rather than sensory deprivation. In the research literature, there has been a trend towards case histories and anecdotes from practitioners' notes, rather than controlled trials with significant patient numbers because of rarity of conditions and because of the individual differences of the losses and impact that these sensory losses may cause.

Recognizing sensory loss can sometimes result in the person functioning above average with the remaining senses. There are numerous anecdotes of compensatory features from the learning disability and autism literature [3] and in cases where children have not been conscious of their loss or difference until later life, such as being born functionally blind, or losing an eye following trauma (accident of disease, cancer), these children may experience heightened aural senses, touch, and olfactory senses [4].

Phantom limb pain has been well documented [5] where patients continue to experience pain and sensations in the amputated limb;

Short Communication

Sensory Compensation in Children Following Vision Loss after Trauma and Disease

Abstract

Sensory compensation or sensory substitution occurs when a sense organ, such as the eye, is lost due to trauma or disease. Individuals often experience phantom limb sensation or pain but research increasingly points towards some individuals developing a heightened level of functioning in their remaining senses, particularly in their remaining intact eye. Losing an eye at an early age can often result in "super functioning" in the remaining eye providing that no similar trauma or disease results. Cases include young children who have undergone enucleation because of diagnosed unilateral retinoblastoma and whose remaining eye is free from disease.

> and those with prostheses often become accustomed to the loss and to the artificial limb [6]. Edmonds [7], suggests differences in ability might be termed "diffability" rather than "disability", emphasizing individual differences instead regarding them as losses.

> Exploration of the impact of health related disorders has opened the gateway not only to acceptance and support, but also to the mechanics behind loss. Anecdotal reports on a loss of sense or senses, suggests that 'super-functioning' may sometimes occur in a person who has lost a sense organ or the functionality of a sensory organ such as the eye or ear. Referred to as 'sensory compensation', this phenomenon is increasingly attracting interest and beginning to be investigated from within a number of different professional disciplines.

> Sensory compensation has also become known as 'sensory substitution' because it can be the replacement of one sensory modality by another, for example, blind individuals may use touch to 'see' [8]. It involves perception but also brain plasticity. Neuroplasticity [9] has been accepted for some time as a process evidenced in recovering stroke patients whose neural brain pathways re-route or re-circuit to find alternative compensatory ways of functioning in the presence of a damaged neural pathway or when function is lost because of an incompletely innervated muscle fiber.

> Reports of compensation by some patients with a loss in sense modality, suggests that a neurological function may be occurring. Karns et al. [10], found that the auditory cortex of individuals born deaf, processes other senses such as touch and vision. Similar results were found by Gougoux et al. [11]. Brain-imaging studies found that the visual cortex in blind individuals was being used for other sense modalities such as touch and smell.

> It is thought that the neural reorganization in the areas of the brain that handles sensory information allows a sensory impaired individual to become progressively sensitive to other sensory stimuli. Typically, when an area of the brain is no longer active, the brain cells die leading to the release of neurotransmitters. Rauschecker [12],



suggests that neurotransmitters reorganize sensory representations that form the neural basis of sensory substitution. With compensation not occurring in every affected individual, it is thought that some neural systems are not plastic and that age and experience plays a key role in this ability.

Neuroplasticity has been found to occur most often in children. This is because our brains are malleable enough at a young age to rewire some circuits that process sensory information. The earlier a sense is lost, the more likely it will be compensated by another sense. Although research into early life compensation is increasing, we are still in an era where this ability is not yet fully understood. Due to the logistics of investigating this theory in humans, the majority of studies have been conducted on animals.

Sensory compensation for loss of vision

Two theories exist to how blindness affects the other senses: (1) blindness may lead to compensation of the other senses; (2) early blindness may halt the development of perceptual and cognitive mechanisms, disallowing the other senses to compensate [13].

Research into cats and ferrets have indicated that early loss of vision is associated with improved ability to localize sounds [14-17] found that cats that were visually deprived for several years as a result of having their eyelids sutured shortly after birth showed increased auditory use in the visual cortex with their auditory spatial tuning being sharpened in the auditory cortex.

Investigating whether compensation occurs in adults, Petrus et al. [18], deprived adult mice of their sight. After one week, the mice were placed in a sound proof chamber where they were subjected to a series of one-note tones to test their hearing. They found that the mice developed more neural connections and were better able to discriminate among pitches and hear softer sounds. Within a few weeks of having their vision restored, their hearing returned back to normal, indicating that the brain is less hard-wired than previously assumed. With mice typically having poor vision but ultrasonic hearing, this finding may not be transferrable to humans. Nonetheless, behavioral and neuroimaging studies have found that blind-sighted humans often demonstrate enhanced auditory abilities [11].

By compensating for their lack of vision by using their other sensory modalities, blind-sighted individuals often outperform sighted individuals in non-visual tasks such as reading Braille [19]; memory retrieval [20]; unfamiliar voice recognition [21]; verbal memory [20]; auditory spatial discrimination [22]; and musical abilities [23]. Interestingly, not all studies have shown this advantage making the theory of neuroplasticity more enigmatic.

Sudden and complete loss of vision results in rapid changes in the visual cortex which reverts back to normal following restoration of sight [24]. Instead of establishing new neural pathways, the visual cortex may be showing abilities which are normally concealed in sighted individuals. Evidence suggests that blind individuals compensate for their lack of vision by having an increased ability to localize sounds [25].

Comparing blind-sighted and sighted individuals, Lessard et al. [8], found that early-blind individuals have superior sound localization

skills particularly when performing monaural localization than binaural localization compared to sighted individuals. This suggests that the visual cortex processes monaural cues more effectively than binaural cues in early loss of sight.

Gougoux et al. [11], found that sighted individuals showed decreased cerebral blood flow in the occipital lobe where the visual cortex is located. Conversely, blind-sighted individuals who perform better on monaural cues were shown to use their occipital areas in this task thus enhancing this ability. Although use of the visual cortex is shown, it is not known if blind-sighted individuals rely on the visual cortex for this ability [11].

Whilst it is natural to assume that these findings are the result of specific brain areas such as the visual and auditory cortex, Striem-Amit et al. [26], found that enhanced capabilities are a result of the task. Using specialized photographic and sound equipment, they found that blind-sighted individuals were able to see and describe objects regardless of sensory modality, visual experience and prior knowledge of the script used. Disparities between and new findings highlight the point that we still do not know how compensation occurs. Various conundrums remain around the theory of neuroplasticity. For example, some individuals have the ability to compensate and others do not; there are differences according to age and experiences; and we still do not have clear and consensual understanding of the neural processes that underlie ability.

Vision with one eye

Research on the effects of monocular deprivation caused by conditions such as strabismus, amblyopia and congenital cataract show negative effects on vision input and functioning; for example, reduced spatial vision capabilities [27]. However, the visual cortex in those who have undergone early life enucleation following retinoblastoma [28]; Thompson et al. [4], has shown compensation for the loss of binocularity [29,30]. These differences point to unilateral enucleation as being a distinct form of monocular visual deprivation [31,32].

Unlike most forms of monocular deprivation where there is some form of visual input, enucleation results in complete and sudden loss of visual input. The age of the individual at the time of loss also plays an important role in outcome with the earlier loss resulting in the sharpening of other senses [30].

Losing binocularity at a young age can result in reduced visual functioning where motion perception is adversely affected [29,30,33] yet some visual spatial abilities are enhanced. These differences in visual functioning are said to be a result of various factors such as neuroplasticity; years of monocular practice following enucleation; and the absence of binocular interactions [30].

Spatial vision is the ability to discriminate spatially defined features. Two primary measures of spatial vision are acuity, which distinguishes details and shapes of objects, and contrast sensitivity which distinguishes an object from its background. Nicholas et al. [34], found that unilateral-enucleated adults had higher contrast sensitivity than controls viewing monocular with those having undergone enucleation before the age of two and performing three-



and-a-half times better. Similarly, unilateral enucleated individuals performed better than normally sighted controls viewing monocular in acuity tests. However, when the controls viewed binocularly their performance equaled the experimental group [35,36].

Motion processing is the ability to judge motion in the visual field. Bowns et al. [37], found that early-enucleated adults and controls have a similar threshold for detecting motion. However, the control group judged the upper visual field as faster, whereas the early enucleated adults found it to be slow. Comparing depth perception, Gonzalez et al. [38], found the those with early enucleation had worse depth perception.

Marotta et al. [39], observed age differences in a reach and grasp task suggesting that over time, unilateral enucleated individuals reduce forward head movements and replace them with lateral and vertical head movement which better placed them for estimating depth. This is because, with one functioning eye, the visual field is reduced by 25% and is not centered on the midline of our body. This results in the individual continually turning their head to look into the blind portion of their visual field [29].

Early-enucleated individuals seem to improve visual performance through learning and due to reorganization of cells responsible for sensory modality [40-44]. During the first few years of life, cells in the visual system form connections that are strengthened by the amount of sensory information received. These connections form visual functions, which can take years to mature. In young individuals requiring enucleation, these connections will be weak and not widely used, thus will atrophy. As the visual system has not reached maturity, reorganization of the cells often takes place to form other connections to our sensory modalities [29].

Moidell et al. [45], found that children enucleated up to four years of age will have neuroplasticity where cell reorganization can occur within hours and involves other sensory modalities [46,47].

Echolocation

Human echolocation is a functional technique used primarily by blind-sighted individuals to navigate and orientate to the environment. By sensing echoes from objects in the environment, blind-sighted individuals can gather spatial information regarding the position, size, material and shape of a sound-reflecting objects [48-50].

Using spatial processing, echolocation is an enhanced ability, which may partly be a result of neural reorganization though little is known regarding the neural basis of echolocation [51,52]. Although many blind-sighted individuals are able to echolocate, individual skills vary; with training, blindfolded sighted individuals can learn to echolocate, though performance is poorer when compared to blind-sighted individuals [49,53,54].

Echolocation was first used by Griffin [55] to describe how bats can navigate in the dark to detect prey by using sound before being studied in other animals such as dolphins and toothed whales [56]. Human echolocators use self-generated sounds such as clicks produced by rapidly moving the tongue behind the teeth [57] or by mechanical means such as tapping a cane against the floor [58].

Echolocation requires three steps: (1) the sound made by the echo locator; (2) the sound and echo superimposed; (3) the echo only [59]. Though audible, the sounds produced contain some strong frequency in the upper part of the range and only last for 10ms [59-61].

In addition to being able to determine objects in their environment, blind-sighted individuals can determine their position in a room. Rosenblum et al. [62], and Wallmeier and Wiegrebe [50], found that blindfolded individuals were able to estimate the distance of objects echo-acoustically with an accuracy of 1m. By using yaw movements of their heads, blind-sighted individuals outperform sighted individuals in echo-acoustic distance discrimination tasks [63].

Echolocation can also help discriminate two-dimensional shapes [64]. Although the majority of these studies focus on a single reflector, Schörnich et al. [60], found that additional reflectors can improve echo-acoustic distance discrimination performance.

Case history

Hughes [65] reported on Daniel Kish. Now 44, Daniel was born with bilateral retinoblastoma, having his first eye removed aged 7 months and his second eye removed aged 13 months. Using echolocation, Daniel is able to navigate through crowded streets, swims, cycles, travels and dances. Our understanding of echolocation has led to the creation of sensory substitution devices for blind-sighted individuals. By emitting a signal with a receiver, the distance between the source and reflecting object is calculated and transformed into an auditory signal, which increases the blind-sighted individual's spatial awareness and independent mobility [65].

Another case history, PT, also diagnosed with retinoblastoma, underwent unilateral enucleation at 9 months, and with the remaining functional eye has exceptional above-average vision. Cases of "supervision" are not uncommon in those who compensate for sensory loss.

Conclusions

Research into compensation has demonstrated that the loss of one sensory modality can be replaced by another, particularly when the loss occurs at an early age [40]. Neuroplasticity can explain the reasons why functionality can continue despite loss of sensory organs, and that new neural connections are strengthened through experience and practice.

Blind-sighted animals and humans have shown enhanced auditory skills, particularly in their ability to locate sounds [18] complete loss of vision in one eye can result in enhanced visual spatial skills but poorer motion processing skills. Whilst brain-imaging studies are aiding our understanding of sensory compensation in humans, we are still years away from fully understanding the neural processes and individual differences that underlie this ability.

Knowledge in this field is providing the opportunity to develop practical devices for individuals with sensory loss, aiding them in an environment that is heavily sensory dependent.

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