Windows into the Visual Brain: New Discoveries About the Visual System, Its Functions, and Implications for Practitioners

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Structured abstract: *Introduction:* In recent years, major progress has been made in understanding the human visual system because of new investigative techniques. These developments often contradict older concepts about visual function. *Methods*: A detailed literature search and interprofessional discussions. *Results*: Recent innovative neurological tests are described that are able to show much more accurately the visual pathways, the process of vision, and the close relationships among sensory modalities. These tests also reveal the remarkable neuroplasticity of the human brain and disorders of connectivity that frequently involve visual function. *Discussion*: How these recent neurological advances may benefit service providers is discussed. *Implications for practitioners*: It is important that from time to time new neurological and ophthalmic developments are summarized for professionals who are involved in the clinical management of individuals with visual disorders and how the newly acquired knowledge affects the diagnosis and intervention strategies. Visual rehabilitation must be based on up-to-date science, which continually changes and grows with research.

Professionals who work with individuals who are visually impaired (that is, those who are blind or have low vision) need to understand recent scientific developments related to vision. Until the late 1800s, vision was attributed entirely to the eyes when it was suggested that the occipital lobes and the eyes together were responsible for seeing. In the mid-1900s, it was proposed that key regions in the brain were responsible for specific visual functions and that these regions were in close proximity to the main visual streams or pathways originating from the occipital lobes. The introduction of investigative neurological tests during the past decade has made it apparent that widespread neuronal networks, rather than specific centers in the brain, are responsible for the

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different visual functions. The discovery of the role of neuronal networks has radically changed thinking about the function of the brain. Regional centers (hubs) are surrounded by a dense network of short connections that collect specific information from highly specialized neurons on certain neurological functions and are further connected to higher-level hubs by longer tracts (Shams & Kim, 2010). Therefore, locally generated specific information from many different areas of the brain is integrated at anatomical regions, but even these regions form networks to synthesize further the different types of perceptual or motor tasks. Clearly, vision, a dominant sense, is closely integrated with other sensory modalities; consequently, the entire brain participates in vision.

Until recently, information about the visual brain came from experiments with animals, human anatomical lesions, histological and biological analyses on postmortem examinations, and studies of various clinical disorders. The introduction of radically new neurological investigative techniques has greatly improved the understanding of the brain. Simultaneously, researchers have begun to superspecialize, which has further increased scientific information. For example, ophthalmologists are able to specialize in retinal disorders, cataracts, glaucoma, visual testing, ocular genetics, and neuro-ophthalmology.

These scientific developments are fragmenting research, but the advances are not filtering down fast enough to practicing medical and educational professionals who provide care to children and adults with visual disorders and who may feel bewildered by the deluge of discoveries. Therefore, the purpose of this article is to describe some of the newly discovered information about the visual system, its functions, and the implications for practitioners.

Neurological tests: Windows into the brain

Professionals who work with individuals with visual impairments should be partially familiar with some of the more commonly used neurological tests. During the 1930s, electroencephalography (EEG) was introduced for the study of electrical activity of the brain to relate it to neurological functions and disorders. Environmental information is transduced into orderly patterns of electrical activity, reflecting the summation of oscillatory frequencies among billions of neurons. Neuronal groups transfer meaningful information by high-frequency oscillations, usually in the gamma-band range of 30–50 in one second. The frequency of oscillations and the interval during which they are generated and perceived are essential. This type of coordinated communication between multiple neuronal groups and the thalamus is required for cognition (Llinás & Ribary, 1993; Llinás, Ribary, Contreras, & Pedroarena, 1998; Whittington, Cunningham, LeBeau, Racca, & Traub, 2011) and is useful in studies of certain functions and disorders (Dobel, Junghofer, & Gruber, 2011). Electrophysiology plays many different roles in the diagnosis of visual disorders (van Genderen et al., 2006).

Magnetic encephalogram (MEG) studies are based on the fact that the electrical currents produced by the brain are always accompanied by magnetic fields. Electrical signals, however, cannot easily pass through the skull but magnetic signals do; thus MEG studies can provide a more detailed analysis. This test is still difficult to conduct with children and, in the field of vision, is used mainly for research (Taylor, Donner, & Pang, 2012).

In computerized tomography (CT scan), the computer combines numerous X-ray images and then creates cross-sectional views of the anatomical structures. Structural magnetic resonance imaging (MRI) also reveals structures, but, in contrast to the CT scan, is not based on radiation. A person lies within a powerful magnet, the MRI scanner, and the magnetic field alters the alignment of tissues in the brain, which then generate radio frequency fields. These changes in tissues can be detected by the scanner, revealing detailed structures with high accuracy. Functional magnetic resonance imaging (fMRI) during certain perceptual or motor tasks or during task-free periods measures the metabolic activity within the various regions of the brain or spinal cord. The test detects changes in blood flow, which are directly related to the consumption of energy. The primary form of fMRI uses blood oxygen level–dependent contrast, but other biomarkers can also be used. The fMRI may be combined with an EEG or near-infrared spectroscopy, but there are also other technical improvements (Lenglet et al., 2012). MRI images have been shown to be useful in the anatomical and functional studies of the visual brain (Bedny, Pascual-Leone, Dodell-Feder, Fedorenko, & Saxe, 2011; Sadato, 2005).

Diffusion tensor imaging tractography (tractography) outlines white-matter neuronal tracts in two- or three-dimensional (hereafter 2- or 3-D) images by using MRI techniques and computer image analysis (Beer, Plank, & Greenlee, 2011; Fernandez-Miranda et al., 2012; Leh, Chakravarty, & Ptito, 2008). The test is based on the fact that within a bundle of normal fiber tract, water diffuses asymmetrically, and the scanner is sensitive to these differences in diffusion. Colors (red, green, blue) are added that show how the fibers are oriented in a 3-D system. Tractography reveals in great detail the extremely complex cross-modal interconnections among the networks that serve different senses, showing that they are not isolated from each other. Both fMRI and tractography have radically improved the understanding of brain function and modified researchers' concepts of neurophysiology.

Near-infrared spectroscopy (NIRS) measures blood oxygenation in the brain. With NIRS, infrared light is shined through the skull, and the pattern of reflected light, which depends on blood oxygenation within the brain, is analyzed.

Positron emission tomography (PET) and single photon emission computed tomography (SPECT) produce 3-D images by detecting gamma rays from injected radioisotopes. Research using PET and SPECT has been applied to the study of visual and other disorders.

Transcranial magnetic stimulation (TMS) of the various anatomical areas has also contributed to improved the understanding of brain functions, including vision. By applying magnetic currents to various regions of the brain, those areas are activated to do certain neurological functions. TMS makes it possible to test the different brain regions in relation to visual, motor, perceptual, and cognitive functions.

Visual pathways

The retinogeniculostriate visual system consists of three major tracts: the dorsal, ventral, and koniocellular pathways that conduct electric messages from the retina to the occipital cortex. The dorsal and ventral streams then forward information toward the frontal lobes. In sighted individuals, the dorsal stream involves the parietal lobes and plays a special role in visual-spatial and movement-oriented tasks. The ventral stream reaches the temporal lobe and participates in visual-perceptual tasks. The koniocellular pathways play a role in the perception of visual movement (Cheong, Tailby, Martin, Levitt, & Solomon, 2011). There are a number of other visual pathways originating from the retina, such as the retino-hypothalamic tracts that are important for circadian rhythm and melatonin production, the tracts for pupilary response, and the collicular (retinal-tectal) visual pathways that are involved in the perception of motion (Lanyon et al., 2009). Readers who are interested in the anatomy of a specific visual pathway should search the Internet under "diagrams of the visual pathways." This approach is more satisfactory than using a single diagram of the brain in the text.

The visual tracts are no longer seen as purely visual because they contain pathways from other sensory modalities. Tractography clearly shows that a large number of pathways are arranged in complex patterns that interconnect all parts of the brain with each other, including the visual tracts. Earlier descriptions of the dorsal and ventral streams are now considered to be inaccurate (de Haan & Cowey, 2011) because of these interrelationships. Even in the occipital cortex, the lateral occipital tracts bring information from the other senses (Fiehler & Rösler, 2010; Kim & Zatorre, 2011). Essential to planning intervention and treatment protocols is the understanding that all neuronal groups in the cerebrum are also connected to the thalamus, which plays a role in consciousness.

Plasticity of the brain

As we discussed earlier, the brain consists of widespread, highly specialized neuronal networks that signal to each other by complex electrical oscillations that are characterized by certain frequencies and patterns (Shams & Kim, 2010). The neurons that are located in the narrow ribbon of cortex (gray matter) over the brain are involved in perceptual tasks, including vision, but more than 90% of the brain contains interconnecting tracts that make up the white matter. Only neurons are responsible for neurological functions, but the interconnecting tracts make the integration of sensory modalities possible. Gray and white matter should always be viewed as functionally inseparable.

The brain is highly sensitive to adverse environments, such as visual impairment or global neglect (Noppeney, 2007), but the human brain also has a remarkable ability to compensate for neurological deficits. The process of these adaptive changes is called neuroplasticity, and this ability, most active in early age, is present throughout life (Shu et al., 2009). For instance, in children with total congenital ocular blindness, the occipital networks that normally mainly serve visual functions are redistributed to process tactile, spatial, and somatosensory information and even language (Watkins et al., 2012). In individuals who are congenitally blind, braille reading by touch involves the occipital visual areas. These neuroplastic changes occur because the occipital regions also receive input from the other senses, but without visual input, the visual neurons are redirected to do other functions.

It is critical to appreciate that neuroplasticity is dependent on age and experience; during early life, when one sense is weaker or absent, the functionality of the other senses can be enhanced, but only when appropriate and meaningful stimulation is provided. Even in adults, continuous learning results in an increase in gray and white matter over the years (Fiehler & Rösler, 2010; Goyal, Hansen, & Blakemore, 2006; May, 2011).

When the retina of persons with severe vision loss that is due to Leber's congenital amaurosis is injected with certain viruses and certain genes, some of the visual functions return (Ashtari et al., 2011). However, there are different age-dependent sensitive periods for the recovery of the various visual functions (Lewis & Maurer, 2005), but they are complex and still not well defined. When children's cataracts are removed, useful recovery depends on multiple factors, such as the onset of visual loss, age at surgery, and whether the children have unilateral or bilateral cataracts. In adults, who have passed their sensitive periods, the correction of congenital ocular blindness may not be appropriate because of their limited neuroplasticity, and this surgery may even be detrimental.

Although genetic factors are mainly responsible for the development of the neurological pathways, the environment also plays a major role. Brain connectivity changes with age as some tracts are strengthened whereas others are cut (pruned). The structural and functional connectivity in individuals with visual impairments *(visual impairment* here refers to disturbed visual acuity, not to other visual disorders) is different from that in individuals who are sighted, but there are also significant differences between those who acquired their visual (ocular and cortical) impairments later in life and those whose blindness or low vision has been present since birth. The touch of a face by an individual who acquired total ocular blindness later in life activates the so-called fusiform area of the temporal lobe and its network. This part of the brain is normally involved in visual face recognition. Another area in the brain is activated, which is again normally involved in the perception of movement, when their arms are touched by a moving object. In contrast, in persons with total congenital blindness, these anatomical regions are not activated; instead, the original visual areas are reassigned to take on entirely different functions (Liu et al., 2007; Sadato, 2005; Watkins et al., 2012; Yu et al., 2008). In conclusion, when blindness is acquired later in life, the well-established visual regions and their networks will perform equivalent perceptual tasks from the other senses (Goyal et al., 2006), but in congenital ocular blindness, many visual networks may cease to exist in time.

There are many more examples that demonstrate the strength of neuroplasticity. When the visual areas in the occipital lobes are destroyed, especially early in life, and the person appears to be without conscious vision, subconscious visual awareness ("blind sight") may still exist because the occipital lobes receive increased visual input from the superior collicular (retinal-tectal) visual pathways (Giaschi et al., 2003; Leh, Johansen-Berg, & Ptito, 2006). People who are blind rely more on vestibular and somatosensory feedback for their mobility than do those who are sighted. The earlier individuals with visual impairments receive special training, the more extensive is the reorganization of their brains that subsequently leads to better coping mechanisms.

Disorders of connectivity

Vision is involved in a vast number of disorders, such as dyslexia, autism, post-traumatic stress syndrome, and many other neurological or psychiatric disorders or syndromes. Synesthesia is a rare disorder of overconnectivity in which one sensory stimulus evokes another unrelated sensory modality (Dovern et al., 2012). Colors may be perceived with taste, music, sounds, letters, numbers, reading, and movement. Reading disabilities can be also be caused by different types of connectivity disorders (Payrin, Démonet, N'Guyen-Morel, Le Bas, & Valdois, 2011; Vandermosten et al., 2012). Thus, the management of dyslexia may need to vary from person to person.

Individuals with misidentification syndromes may claim that their body parts do not belong to them or may even perceive that their close relatives are imposters (Hirstein, 2010). Alice-in-Wonderland syndrome is characterized by temporary distortions of visual and auditory perception, changes in the concept of time and body image, and even hallucinations and déjà-vu experiences (Brumm, Walenski, Haist, Robbins, Granet, & Love, 2010). Because of a viral illness or genetic or other factors, the connections between various networks are temporarily disturbed.

Prosopagnosia (difficulty in recognizing faces) is a relatively common connectivity disorder that has different causes. It is often claimed that the fusiform area of the temporal lobe is responsible for facial recognition, but, because it is only one of the centers in a vast network of connecting tracts, it would be more appropriate to state that this area plays an important integrating role. Furthermore, the prefrontal cortex is responsible for final facial recognition, deciding that a face is familiar or not (de Borst et al., 2011; Rapcsak & Edmonds 2011). Obviously, these connectivity disorders require entirely different management.

Discussion and implications for practitioners

It is important to note that many earlier concepts about brain functions were incorrect, which is not surprising because of the extreme complexity of the central nervous system. However, several conclusions can be drawn from recent advances that would benefit professionals who are providing a variety of services to children and adults with visual disorders. Connectivity studies have clearly proved that vision is more complex than was previously thought, and it is not isolated from the other senses but works in close liaison with them. The brain is plastic at any age, but remarkably so during the early years of life. Therefore, exposure to meaningful experiences and age-dependent activities will not only strengthen the visual networks, but will modify the other senses in compensation for the loss of vision.

Recently introduced innovative tests will benefit individuals with visual impairments who have disturbed sensory modalities. For example, tractography studies will be able to estimate the network capacity of the brain, and when the sensory exposure is too great for that person, tactile defensiveness, sensory overload, and an inability to perceive information from different senses simultaneously may occur (Wozniak et al., 2013). Also, information obtained from the innovative neurological tests may improve the quality of intervention services secondary to an improved understanding of the nature of the neurological structures involved. In children who were initially assumed to have significant visual impairments, EEG, CT, and MRI technologies were able to identify the cause of their severe visual inattentiveness as being due to dyskinetic eye movements, rather than the loss of visual acuity (Jan, Lyons, Heaven, & Matsuba, 2001). Diagnostic information yielded by these tests resulted in more appropriate intervention strategies.

Research has shown that in young otherwise typical children who have low vision because of ocular deficits, the promotion of visual activities can quickly increase the function of cortical visual areas, whereas when children are totally blind, developmentally appropriate, carefully designed meaningful learning environments will strengthen the other sensory modalities. In contrast, on the basis of years of clinical and empirical experience, service providers have found that the learning environment must be tailored carefully to children who have ocular or cortical visual impairments with multiple other disabilities because it may lead to sensory overload and behavioral consequences (Jan, Sykanda, & Groenveld, 1990). It is reassuring to know that professionals who have been working with children with visual impairments understood these therapeutic principles many years before research proved them to be correct.

It is important to identify visual loss as early as possible and to assess functional vision, regardless of children's diagnoses, learning competence, and needs, to ensure prompt and appropriate early intervention for children with low vision, blindness, and cortical visual impairment, and to emphasize the importance of individual variability and the process of neuroplasticity for growth at all ages. Ocular disorders in early childhood may adversely influence visual development. Even in healthy adults, experimentally induced myopia shows a marked reduction of fMRI activity in the visual cortex (Mirzajani, Vilayphonh, Vasseur, Caputo, Laloum, & Chokron, 2011). Ophthalmologists know that myopia in early childhood must be corrected because it leads to disordered visual development in addition to changes to other sensory modalities. The brains of children who have visual and other developmental difficulties do not warn them about their deficiencies. Only when these children begin to socialize with other children do they realize that they are different, which may lead to behavioral difficulties. Early comprehensive assessments are important for children with visual disorders, and services must be planned in such a way that the development of all the other sensory modalities is promoted. Appropriate services for children with multiple disabilities must be integrated and based on the entire child, not just on a single area of challenge. For example, the visual benefits of corrective eyeglasses in a child with multiple disabilities are greater following strengthened neck control. The rehabilitation should begin after the early diagnosis of visual and other deficits, and the sooner the better.

In the field of health care, there is a growing tendency to rely more on tests and less on clinical evaluations. However, sophisticated imaging and other studies, while offering much information about the workings of the brain, cannot replace functional evaluations of individuals with visual disorders within and across meaningful contexts and environments. At this stage, these tests do not accurately reveal function or developmental potentials. For example, how children with cortical visual impairment function in school, at home, and in other environments must be carefully evaluated by experienced professionals before educational and other services can be organized. Such evaluations are essential because these children not only have disturbed visual acuity and field loss because of fewer functioning neurons in the occipital cortex (the main characteristic of this visual disorder), but almost invariably have various additional neurological deficits that interact in complex ways. Therefore, each child has diverse and unique needs and requires corresponding intervention strategies.

A classification system that emphasizes single anatomical regions for neurological tasks is misleading because widespread networks are responsible for even the simplest functions. Therefore, visual conditions should be carefully defined and classified by their functional characteristics or by a combination of functional and network abnormalities. Facial recognition difficulties, cortical visual impairment, and dyslexia and disorders of eye movements and visual attention may all be the result of highly variable causes and network abnormalities. It has been repeatedly shown that careful definitions of disorders and their classifications are critical for the delivery of appropriate services. Identifying vastly different visual conditions simply as cerebral visual impairment is confusing and harmful. Furthermore, the visual networks and their functions of sighted individuals and individuals with low vision or blindness should not be equated because they are not identical.

Recent research has increased knowledge of the nature of a vast number of neurological and developmental conditions in which vision plays a major role. It is not possible for one professional to provide intervention for individuals with all these complex disorders because these individuals need different and highly specialized services. As always, good communication and teamwork are required between service providers and the involved families.

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