

Blindness

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Blindness is the complete absence or severe impoverishment of vision. The World Health Organization defines blindness as acuity of less than 20/400 in the better eye (in case of unequal vision loss across the two eyes) or a visual field of less than 10 degrees around fixation.

Blindness is to be distinguished from 'low-vision' which is defined as best-corrected acuity of less than 20/60, but better than 20/400. While blindness effectively renders vision ineffective for even the most basic tasks such as obstacle avoidance, low-vision permits some coarse visually-guided skills although it compromises more detail-oriented ones such as reading and driving.

Over 35 million people across the world are blind, with the highest incidence (over 90%) in developing countries. Each year, 1 million blind people get treated, 6 million people die blind, and 8 million new cases are added. Thus, the net yearly increase of the blind population is 1 million. The economic cost corresponding to the loss of productivity caused by blindness is estimated to be in excess of \$40 billion per year.

Primary causes of blindness:

Blindness can have a variety of etiologies ranging from damage to the eye to brain-trauma. The causes that account for the bulk of global blindness are:

Cataracts: A cataract is an opacity of the lens. The likelihood of an opacity increases with age. Although the precise causal factors are still not clearly understood, UV exposure, genetic predisposers, smoking and diabetes mellitus are all believed to heighten the risk. Cataract surgery is one of the great success stories of our time. A few-minutes long procedure that involves removal of the lens done under local anaesthesia (for adults) can almost completely restore clear vision.

Macular degeneration (MD): The macula is the central region of the retina that includes the fovea and which subserves high-acuity vision. MD is a disorder that compromises the macula and thus severely impairs visual perception. The risk for MD worsens with age and prior familial incidence of the condition. Regrettably, no satisfactory preventative or treatment strategy for MD is currently available.

Diabetic retinopathy: Overgrowth and/or hemorrhaging of retinal blood-vessels associated with diabetes mellitus can lead to retinal damage and eventually, blindness. Effective interventions exist, including laser based cauterization of hemorrhaging vessels.

Glaucoma: Glaucoma is chronically increased intra-ocular pressure, which can lead to damage of the retinal tissue and, especially, the optic disk. Glaucoma might have genetic underpinnings in some individuals, and might be the outcome of trauma in others. Treatments that reduce intra-ocular pressure are effective in the early stages of glaucoma. Delay leads to permanent damage to the optic nerve fibers, and hence uncorrectable visual loss.

River-blindness (onchocerciasis): Onchocerciasis is a common cause of blindness in parts of Africa. The disease is caused by a skin-dwelling nematode whose larvae are transmitted across a population via biting blackflies. The flies breed in running streams (hence the prevalence of the disease near rivers). The transmitted larvae migrate across a person's body. In the eyes, these larvae cause a host of problems including corneal scarring, glaucoma and cataracts, leading eventually to blindness. Population-based drug-distribution programs have been effective in reducing the incidence of onchocerciasis.

Factors that are implicated in childhood blindness include maternal rubella (infection during the first trimester of pregnancy greatly increases the likelihood of blindness in the child), measles (which causes corneal scarring), vitamin A deficiency (which can lead to corneal drying and scarring), conjunctival infections at birth, and retinal detachment associated with premature births.

Based on an analysis of causal factors, it is believed that nearly three-quarters of all blindness in the world is either preventable or treatable. Large-scale programs, such as WHO's Vision 2020, have been launched to reduce and eventually eliminate all cases of avoidable blindness within the first few decades of the 21st century.

Scientific studies of perception in the blind:

The study of blindness provides a unique opportunity to investigate diverse aspects of sensory/perceptual processing as well as issues related to brain plasticity and learning. We briefly describe a few representative studies below.

Blindness induced changes in non-visual perceptual sensitivities: How accurate is the popular belief that the blind can 'hear better' than the sighted? Surprisingly, there have been no definitive answers to this question. Several studies have failed to find a consistent enhancement of auditory sensitivities in the blind relative to the sighted. However, a recent report from Pascal Belin and colleagues at the Montreal Neurological Institute shows that when the performance of early blind individuals (blindness onset within the first two years of life) is examined separately from that of late-blind ones, an advantage in musical note perception becomes evident for the former. Late-blind subjects show no such improvement relative to the normally-sighted. This result and others like it highlight the plasticity of perceptual processing mechanisms of the brain and also suggest that the initial years after birth might be especially amenable to such neural resource reorganization.

The importance of visual input during the first few months of life: How do the visual abilities of individuals who have been blind for the first few months of their lives and have then gained sight, differ from those of their normally sighted counterparts? Answering this question is important for understanding the processes of early visual learning, and time-constraints on neural plasticity. Daphne Maurer and her colleagues have systematically studied several children who were born with congenital cataracts and were treated within the first half-year after birth. Interestingly, when these children are tested several years after the surgery, they exhibit subtle but consistent impairments in their ability to extract configural information from images for tasks such as face recognition. These results suggest that early visual information is important for

organizing the perceptual machinery needed to properly analyze subtle spatial relationships in images. Exactly how the early visual input contributes to the development of this spatial perception ability remains unclear.

Language learning by blind children: What role does visual information play in bootstrapping the development of linguistic abilities in children? A strong case can be made for vision as a critical modality for grounding linguistic constructs in real-world entities. The use of prepositions and the learning of nouns, for instance, is expected to be greatly facilitated by visual information. Is this really true? A powerful way of addressing this question is to look at language learning in congenitally blind children, and to examine whether and how early language in the blind differs from that in the normally sighted cohort. Pioneering studies along these lines were undertaken by Barbara Landau and Lila Gleitman. Their work and subsequent investigations have demonstrated an impressively intact facility for language learning in the blind although the acquisition of some entities that do not have immediate sensory counterparts shows a delay. At the very least, these studies demonstrate that visual information is not a crucial building block for basic language learning. In future work, it will be interesting to explore what role, if any, vision plays in specific and more subtle aspects of linguistic skills.

The feasibility and mechanisms of late visual learning: The classical notion of ‘critical periods’ suggests that visual learning is possible primarily during the early stages of development. Just how strict is the critical period for visual learning? Is all visual skill acquisition permanently compromised if the brain is deprived of input from the eyes for the first few years of life? Early animal studies of dark-rearing have indicated that early visual deprivation has dramatic consequences on subsequent development. The subjects in these studies exhibited profound and permanent deficits in visual development, as would be predicted by the ‘critical periods’ idea. Does this apply to humans as well? This question has been difficult to answer since cases of sight onset late in life have been very rare. However, a new initiative launched by Pawan Sinha of MIT, Project Prakash, is allowing a systematic investigation of this issue. The effort identifies congenitally blind children in India and provides them treatment, thus fulfilling an important humanitarian need. But, in doing so, it also affords an opportunity to scientists to examine how much visual skill acquisition is possible after several years of blindness. The results so far

suggest that the strict notion of critical periods needs to be refined. The treated children are able to acquire several complex visual skills within several months of gaining sight. Longitudinal studies of the skill acquisition process have provided insights into the process by which visual learning proceeds in these children, and perhaps, in normal development. Planned neuro-imaging studies will allow for a structure-function analysis by revealing which neural activations are correlated with the onset of different behaviorally observed skills.

Cortical reorganization in the blind: What happens to parts of the brain that are deafferented, i.e. cut-off from inputs? This is a question of direct import for our understanding of brain plasticity and functional re-organization. Neuro-imaging studies of blind individuals provide an excellent way of addressing this issue. In the normal human brain, over 30% of the cerebral cortex is devoted to processing visual information. What happens to this cortical tissue in the brain of a congenitally blind person? Does it sit silent and unused, or is it recruited by the brain for alternative functions? Is there an age beyond which such recruitment cannot be achieved? These and related questions are being addressed using techniques such as functional magnetic resonance imaging. The results so far have been very interesting. The visual cortex in blind subjects has been reported to exhibit activations corresponding to tactile stimulation as well as linguistic tasks. For instance, touching Braille letters leads to visual cortical activation in the blind. Interestingly, data suggest that these activations are likely to be functionally significant. Temporary disruption of the primary visual cortex via trans-cranial magnetic stimulation decreases Braille reading performance of blind subjects but not that of blindfolded sighted ones.

Cross-talk between sensory modalities: We regularly experience the world via multiple sensory modalities. A ball feels smooth to the touch and looks round. A cube has sharp edges in both modalities. A long-standing question that philosophers and brain scientists alike have grappled with is this: Do the different modalities contribute to an ‘amodal’ description of the world, or are their representations modality specific? This issue has been called the ‘Molyneux query’ after a letter Molyneux wrote to the English philosopher John Locke. The query has resisted an answer for over three centuries. An answer will be very significant not only from the basic science perspective, but also from the applied viewpoint of designing sensory-substitution devices to help the blind. Will the blind be able to interpret visual information if it is translated into a

different sensory modality? We do not yet have a definitive answer, but with ongoing studies of people who gained sight late in life, we do now have a chance of resolving this question.

Frontiers in blindness research:

Blindness is one of the most debilitating handicaps. It impacts all aspects of a person's life spanning social interactions, education, and career choices. For the many blind individuals in the developing world, the situation is dire, with significantly reduced prospects for health and longevity. Several avenues of research need to be pursued to lessen the global incidence of blindness. Medical research is needed to search for treatments for conditions that currently are incurable. Epidemiological research along with novel service delivery methodologies are needed to identify people in need of treatment and bring them in the fold of medical care. Finally, research into assistive devices is needed to develop aids for the permanently blind. Devices already in the pipeline include 'smart canes' that use range-sensors and vibro-tactile actuators to alert blind users of obstacles in their path, GPS augmented way-finding systems, and retinal/cortical implants to compensate for missing retinal tissue or optic nerve damage. Also under development are systems for the partially sighted that can enable people to most effectively utilize the residual vision that they possess. These are exciting scientific enterprises, made all the more gratifying because they carry with them the promise of improving the lives of millions of blind individuals across the world.

Suggested readings:

Gougoux, F., Lepore, F., Lassonde, M., Voss, P., Zatorre, R. J., Belin, P. 2004, Pitch discrimination in the early blind. *Nature*, 430, 309.

Landau, B. and Gleitman, L.R. 1985, *The development of speech in blind children. Language and Experience: Evidence from the Blind Child.* Harvard University Press.

Le Grand, R., Mondloch, C. J., Maurer, D. and Brent, H. 2001, Early visual experience and face processing. *Nature*, 410, 890.

Mandavilli, A. 2006, Look and Learn. *Nature*, 441, 271-272.

Morgan, M.J., 1977, Molyneux's Question: Vision, Touch and the Philosophy of Perception, Cambridge: Cambridge University Press.

Sadato, N., Pascual-Leone, A., Grafman, J., Ibanez, V., Deiber, M.-P., Dold, G. and Hallett, M. 1996, Activation of the primary visual cortex by Braille reading in blind subjects. *Nature*. 380: p. 526-528.

Bouvier, J. V. and Sinha, P. (2007). Object concept learning: Observations in congenitally blind children and a computational model. *Neurocomputing*. Vol. 70, 2218-2233.

Ostrovsky, Y., Andalman, A. and Sinha, P. (2006). Vision following extended congenital blindness. *Psychological Science*. Vol. 17, No. 12, 1009-1014.