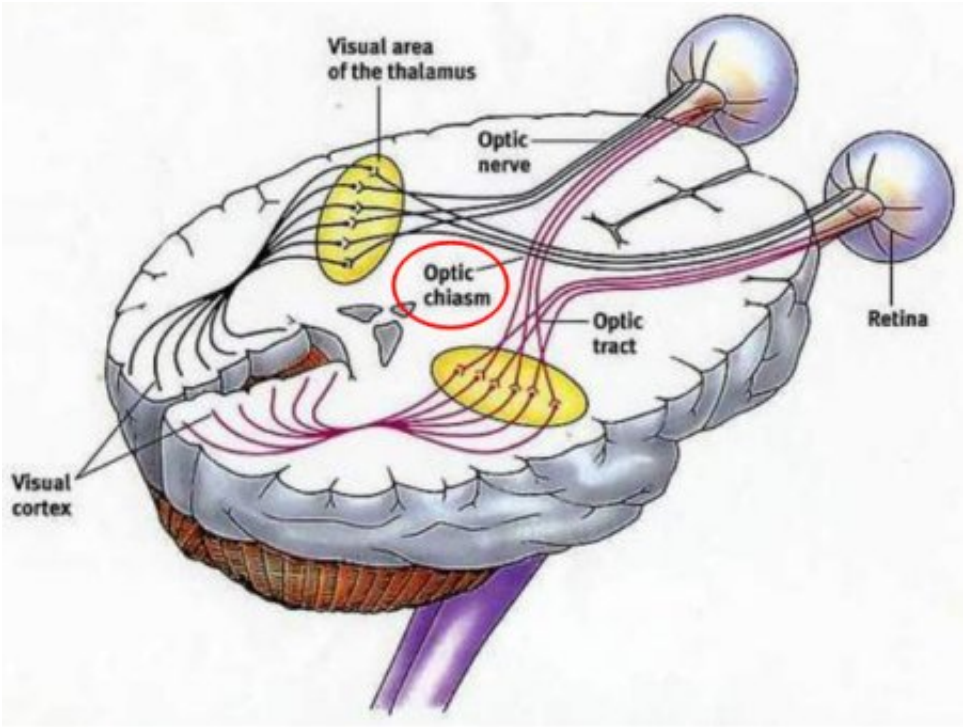
# Congenitally absent optic chiasm: Making sense of visual pathways

April 15 2013, by John Hewitt

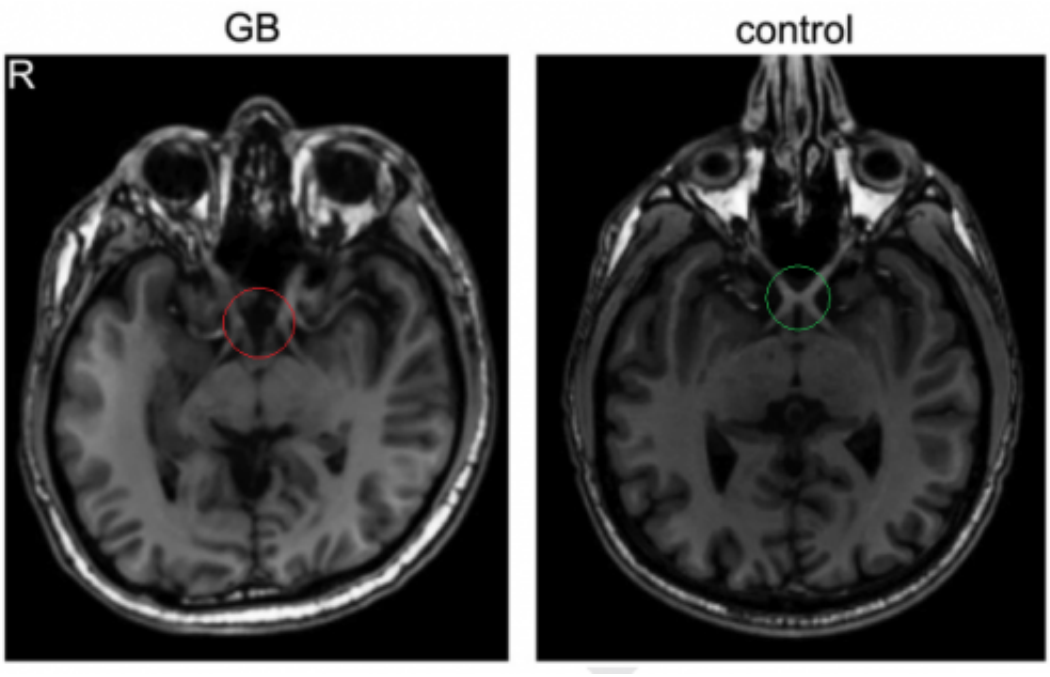



Optic Chiasm. Credit: glaucoma-eye-info.com

(Medical Xpress)—One way to increase our understanding of bilateral brains, like our own, is to inspect their paired sensory systems. In our visual system, the optic nerves normally combine at a place called the optic chiasm. Here half the fibers from each eye cross over to the opposite hemisphere. When this natural partition fails to develop normally, the system compensates in different ways. In people with

albinism, for example, almost all of the fibers fully cross at the chiasm. As a result, images are combined in the brain in such a way that full depth of vision is limited. Their eyes also may move slightly independent of each other, or dart back and forth in a condition known as nystagmus. When the opposite situation occurs, that in which the optic nerves do not cross at all during their development, it is called congenital achiasma. An individual with this rare condition was recently studied with different forms MRI. The results, reported in the journal *Neuropsychologia*, show that achiasma can occur as an isolated defect, lacking any structural abnormalities in other pathways that cross the midline. The study also demonstrated that the part of the cortex that first receives the visual input, the primary visual cortex, does not rely on information from the opposite side to perform its immediate functions.

When input to the two halves of the brain is parsed according to the eye rather than to the visual field, binocularity is typically affected in some way or another. The eyes may have a slightly crossed configuration, and nystagmus occurs more readily as the visual system updates. The subject of the present study, henceforth known as GB, additionally displayed an eye effect known as seesaw nystagmus. In this type of nystagmus, the eyes alternately move up and down, out of phase with each other. When initial MRI scans failed to show an optic chiasm in patient GB, researchers subsequently verified that it was completely absent by tracing the nerves with diffusion tensor imaging (DTI). The subject was also given a series of tests during a functional MRI scan (fMRI) in order to see how the visual field mapped to his cortex.




Structural T1-weighted MRI scan shows the optic chiasm in a control subject but not in GB. Credit: *Neuropsychologia*, DOI: 10.1016/j.neuropsychologia.2013.03.014

By dividing the visual field into four quadrants, and presenting a stimulus to each in turn, the researchers confirmed their suspicions that each hemisphere was mapping the whole visual field. To the level of detail available from the MRI scans, both halves of the visual field, the nasal and temporal retinal maps, were found to overlap completely. The researchers also showed that in the primary visual cortex, monocular stimulation activated only the ipsilateral (same side) cortex. Higher cortical areas, such as the V5 motion-associated area, and the fusiform face region, could be activated binocularly.

The MRI scans further showed that the all parts of the corpus callosum, including those that connect the visual cortex, were intact and of normal size. It appears that at the level of V5 and above, the callosum

contributes significantly to binocular integration. In a normal brain, with a normal chiasma, callosal projections connecting the primary visual cortex might also contribute to the seamless integration of the visual scene across the midline. For rapidly moving objects however, it is unclear how the signal delays introduced by the comparatively long fibers that cross the hemisphere would be handled. Alternatively, these projections may be more involved with attention, or with more complex effects like binocular rivalry.

It is still not entirely known why the chiasma occasionally fails to develop. The condition can be genetic, but probably also involves factors like conditions inside the womb. Animal models have demonstrated the effects of various extracellular matrix and cell adhesion molecules on chiasma development. Specifically, axon guidance has been shown to be regulated by the expression of molecules such as NR-CAM, neurofascin, and Vax-1. While a deficiency in any one of these molecules can have effects on the chiasma, any effects must be considered in context of a much larger puzzle. Vax-1, for example, can cause complete absence of the chiasma, but it is also accompanied by various other midline anomalies. These include problems with the development of the callosum, something not seen here with patient GB.

The source of binocular activation of motion and object-specific areas in GB is also a point of interest. There are many channels through which this activation could occur, including indirect projections from subcortical regions involved in visual processing. Further study of patients like GB, together with more detailed genetic information about them, will help us understand how the visual system develops, and how the visual world integrates within a bilateral mind. Once we can do that, perhaps then we will be able to explain other unique cases, like for example, the woman who sees everything upside down.

**More information:** Functional organisation of visual pathways in a

patient with no optic chiasm, *Neuropsychologia*, dx.doi.org/10.1016/j.neuropsychologia.2013.03.014

**Abstract**

Congenital achiasma offers a rare opportunity to study reorganization and inter-hemispheric communication in the face of anomalous inputs to striate cortex. We report neuroimaging studies of a patient with seesaw nystagmus, achiasma, and full visual fields. The subject underwent structural magnetic resonance imaging (MRI), diffusion tensor imaging (DTI) studies, and functional MRI (fMRI) using monocular stimulation with checkerboards, motion, objects and faces, as well as a retinotopic quadrantic mapping. Structural MRI confirmed the absence of an optic chiasm, which was corroborated by DTI tractography. Lack of a functioning decussation was confirmed by fMRI that showed activation of only ipsilateral medial occipital cortex by monocular stimulation. The corpus callosum was normal in size and anterior and posterior commissures were identifiable. In terms of the hierarchy of visual areas, V5 was the lowest level region in the hierarchy to be activated binocularly, as were regions in the fusiform gyri responding to faces and objects. The retinotopic organization of striate cortex was studied with quadrantic stimulation. This showed that, in support of recent findings, rather than projecting to an ectopic location contiguous with the normal retinotopic map of the ipsilateral temporal hemi-retina, the nasal hemi-retina's representation overlapped that of the temporal hemi-retina. These findings show that congenital achiasma can be an isolated midline crossing defect, that information transfer does not occur in early occipital cortex but at intermediate and higher levels of the visual hierarchy, and that the functional reorganisation of striate cortex in this condition is consistent with normal axon guidance by a chemoaffinity gradient.