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# The Visual Pathway—Functional Anatomy and Pathology

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**Visual failure of any kind is a common clinical presentation and indication for neuroimaging. Monocular deficits should concentrate the search to the anterior (prechiasmatic) visual pathway. Bitemporal hemianopia suggests a chiasmatic cause, whereas retrochiasmatic lesions characteristically cause homonymous hemianopic defects. Quadrantanopias usually arise from lesions in the optic radiations. Disorders of visual perception can be broadly divided into "where" and "what" problems caused by lesions in the parietal and temporal lobes, respectively, and their associated white matter tracts. Visualization of the retrochiasmatic visual and visual association pathways is aided by diffusion tensor imaging.**
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## Introduction

An understanding of the anatomy of visual pathways is fundamental to the interpretation of imaging performed in the investigation of visual failure and visual field defects. The functional anatomical components of the central visual pathways from the optic nerves to the higher cortical centers have been considered. The focus is on the presenting visual complaint and subsequent lesion localization, as this mirrors how such problems are encountered and approached in clinical practice. This review does not provide an exhaustive list of the various pathologies that can afflict the visual pathways, rather it serves as a framework on which to base a systematic review of the visual system in the context of visual deficit.

## Monocular Blindness

Lesions affecting the retina or optic nerve result in some degree of ipsilateral field defect. The axons of the retinal ganglion cells pierce the sclera of the globes to form the optic nerves. These pass into the skull from the orbits via the optic canals of the sphenoid bone. The optic nerves enter the middle cranial fossa, and at a point anterior to the infundibulum of the pituitary gland, the medial fibers decussate to form the optic chiasm.[1] The optic nerve can be divided into intraocular, intraorbital, intracanalicular, and intracranial (cisternal) parts.[2] Optic nerve pathology can be divided into intrinsic and extrinsic lesions.

Intrinsic lesions of the optic nerve include optic neuritis (ON) and ischemic optic neuropathy (ION). ON refers to the inflammation of the optic nerve and may be anterior, papillitis, with disc swelling, or retrobulbar in which case the optic disc appears normal. It is usually a primary demyelinating process and may occur in isolation or in association with multiple sclerosis (MS) or other demyelinating conditions.[3] The cardinal symptoms of ON are loss of visual acuity and pain in the affected eye, both of which are reported in greater than 90% of patients.[4] The visual field defect has classically been described as a central one, although almost any type of defect may occur.[3] Dyschromatopsia (loss of color vision) may also be present in approximately 90% of cases;[5] it is unusual to have poor acuity and maintain color vision.[6] Equally, dyschromatopsia with only mild to moderate loss of acuity is sensitive for ON.[7] These features help differentiate ON from other intrinsic lesions such as ION, which is typically painless[8]; dyschromatopsia, when present, is usually proportionate to the loss of visual acuity.[9] The imaging findings of ON classically include enhancement of an enlarged optic nerve on fat-suppressed T1-weighted magnetic resonance (MR) images and high signal on fat-suppressed T2-weighted sequences such as short tau inversion recovery (Fig. 1).[10] ON remains a clinical diagnosis, and the main role of MR imaging (MRI) is in the identification of white matter lesions and prognosis for MS.[11] There has been some work suggesting that diffusion tensor imaging (DTI) parameters correlate with visual

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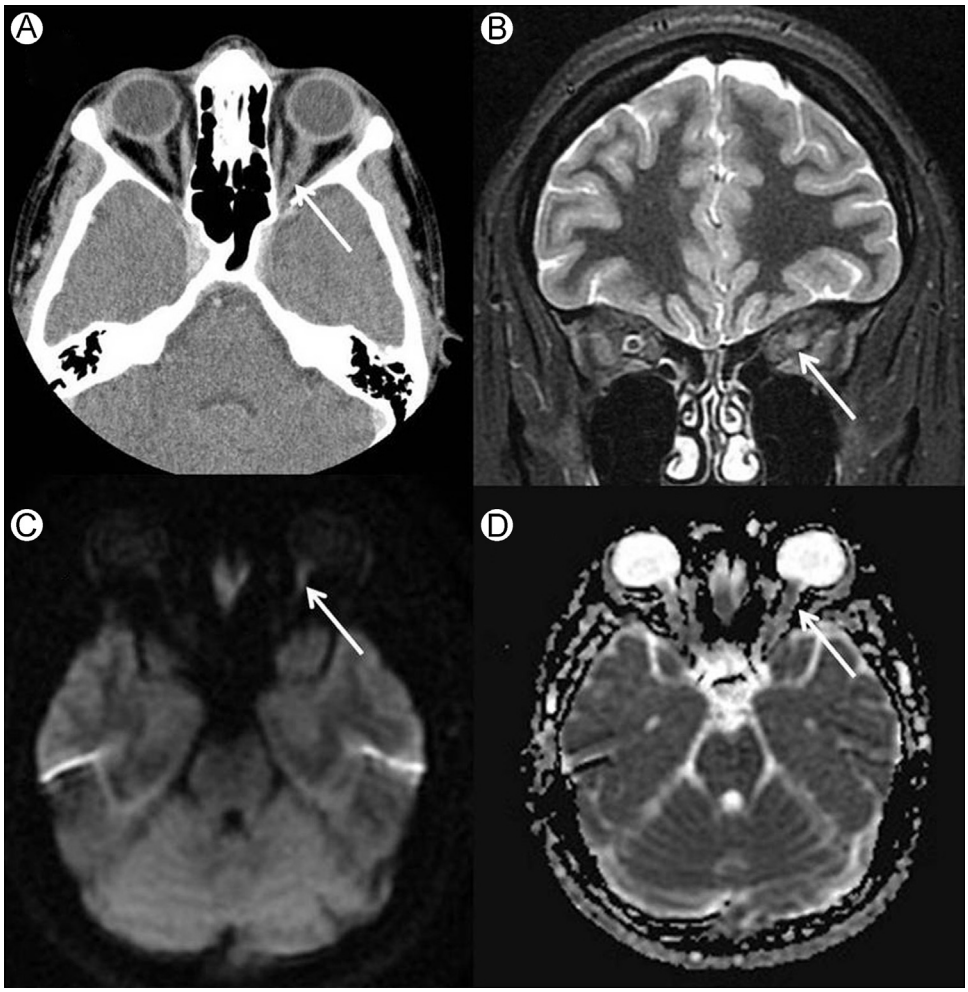


**Figure 1** Optic neuritis: (A) axial CT showing mild left optic nerve enlargement (arrow, compare with right); (B) coronal STIR image showing enlarged left optic nerve (arrow) with effacement of surrounding CSF in the nerve sheath (compare with right); (C) axial DWI; and (D) ADC map. In acute cases there is evidence of mild diffusion restriction (arrows, C and D). ADC, apparent diffusion coefficient; CSF, cerebrospinal fluid; CT, computed tomography; DWI, diffusion-weighted imaging; STIR, short tau inversion recovery.

impairment and axonal integrity and may predict visual outcomes, although studies are limited.[12-15]

ION is a collective term for a number of ischemic syndromes of the optic nerve and is usually subdivided into anterior ION, affecting the optic nerve head, and posterior ION, which involves the remaining 3 portions of the optic nerve.[9] Anterior ION is the commonest entity and presents with sudden-onset painless monocular visual loss, often noticed on waking, typically in an inferior altitudinal pattern.[9,16] Diffusion restriction within the optic nerve has been described in case reports often in the context of posterior ION as a perioperative complication.[17-22] Additional findings include high signal on T2 and fluid-attenuated inversion recovery (FLAIR) and contrast enhancement of the optic nerve or nerve sheath on MRI,[19,22,23] although the overall abnormalities seen are less common in ION than ON.[24] Diffusion restriction is also reported in ON, meaning that the distinction with ION relies primarily on clinical factors.[17] Tractography has been cited as a potential tool in predicting visual recovery,[20] although further studies are needed.

Compressive optic neuropathy (CON) affecting the prechiasmatic optic nerve may result from many different extrinsic compressive lesions, and clinical findings can guide localization. The range of pathologies includes neoplastic, inflammatory, infective, and vascular entities.[25] Lesions causing swelling of the optic disc, usually within the orbit, optic canal and, less frequently, intracranial portion of the optic nerve, constitute anterior CON. However, most cases of CON are not associated with optic disc swelling.[26] Other presenting symptoms of orbital tumors include proptosis, ophthalmoplegia, congestion, and visual loss, although this may be subtle, gaze dependent, and limited to color vision.[25,26] CON without optic disc swelling, that is, retrobulbar CON, may present insidiously with painless, slowly progressive visual symptoms. Patients may also have blurring or "dimming" of vision, and color vision may also be affected, as in anterior CON.[25] The slow onset differentiates most of the cases of CON from ON or ION but causes such as pituitary apoplexy or arterial aneurysm may lead to sudden visual loss.[27,28] Both computed tomography and MRI have a role in the assessment of CON. MRI has superior soft tissue contrast (Fig. 2) and can demonstrate the presence or extent of lesions beyond the orbital apex. However, computed tomography has a role in assessing calcification (Fig. 3)

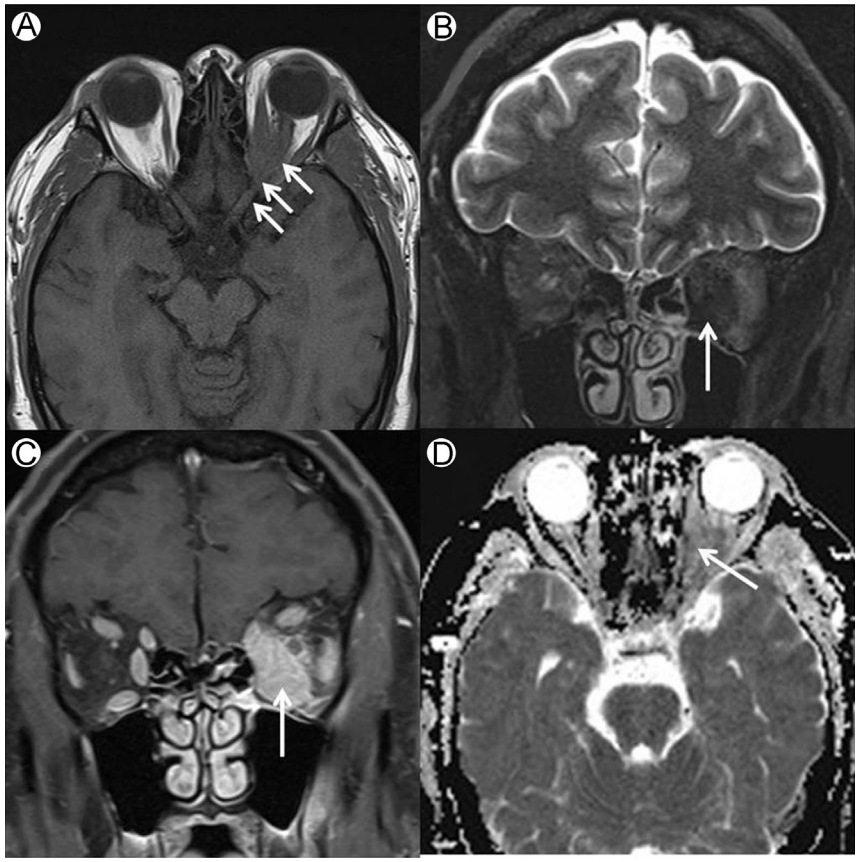


**Figure 2** Compressive optic neuropathy (CON): (A) axial T1-weighted; (B) coronal STIR; (C) coronal T1-fat-saturation postcontrast; and (D) axial DWI images of a transpatial, solid enhancing lesion that shows diffusion restriction (arrows, B-D). The optic nerve is encased and compressed (arrows, A). Biopsy revealed diffuse large B cell lymphoma. DWI, diffusion-weighted imaging; STIR, short tau inversion recovery.

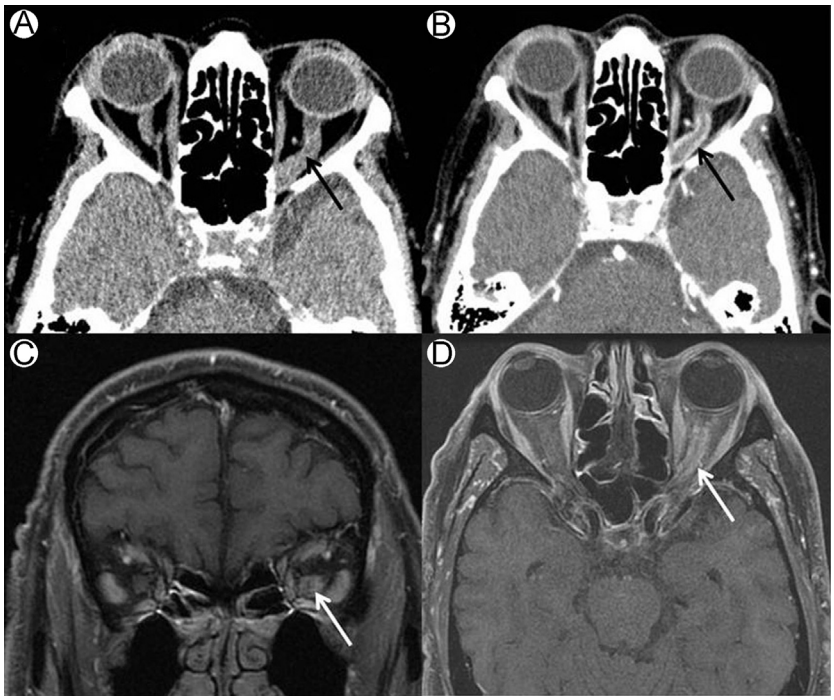


**Figure 3** Left optic nerve sheath meningioma: (A) precontrast CT scan shows patchy high-density calcification (arrows) and (B) postcontrast CT shows tram-track enhancement (arrows) confirmed on the coronal and axial T1-fat-saturation postcontrast MRI (arrows, C and D). CT, computed tomography.

and osseous changes and is quicker and frequently more accessible than MRI.[25,29]

## Bitemporal Hemianopia

Lesions in the region of the optic chiasm can cause a variety of visual symptoms owing to the conformation of the nerve fibers; the characteristic defect is that of a bitemporal hemianopia. The intracranial portions of the optic canals open into the chiasmatic sulcus superoanterior to the ridge of the tuberculum sellae.[30] Here, or just posterior, the medial fibers of the optic nerves (containing visual information from the temporal fields) decussate to form the optic chiasm. The lateral fibers, containing information about the nasal visual fields, do not cross. Behind the chiasm, each optic tract transmits information regarding the contralateral hemifield of vision. The optic tracts continue posterolaterally with most fibers passing around and behind the tuber cinereum and anterior perforated substance and around the cerebral peduncles to terminate in the lateral geniculate nuclei (LGN) of the thalami.[30,31] There are also connections from the optic tracts to the superior colliculi and pretectum of the midbrain concerned with saccadic eye movements and pupillary reflexes respectively.[32]

Lesions around the chiasm can, somewhat artificially, be divided into those affecting the anterior angle, the body, and the posterior angle.[33] Anterior lesions produce varying degrees of temporal field defect in the ipsilateral eye through compression of medial (nasal retinal) fibers, although monocular blindness is possible if the lesion is extensive enough. Contralateral superior temporal field defects ("junctional scotoma") have been reported from anterior chiasmatic lesions.[34] The controversial but proposed mechanism is through involvement of "Wilbrand knee," where already crossed inferonasal fibers loop into the contralateral optic nerve before continuing in the optic tract. However, some contest its existence, believing it to be an artifact secondary to enucleation.[35,36]

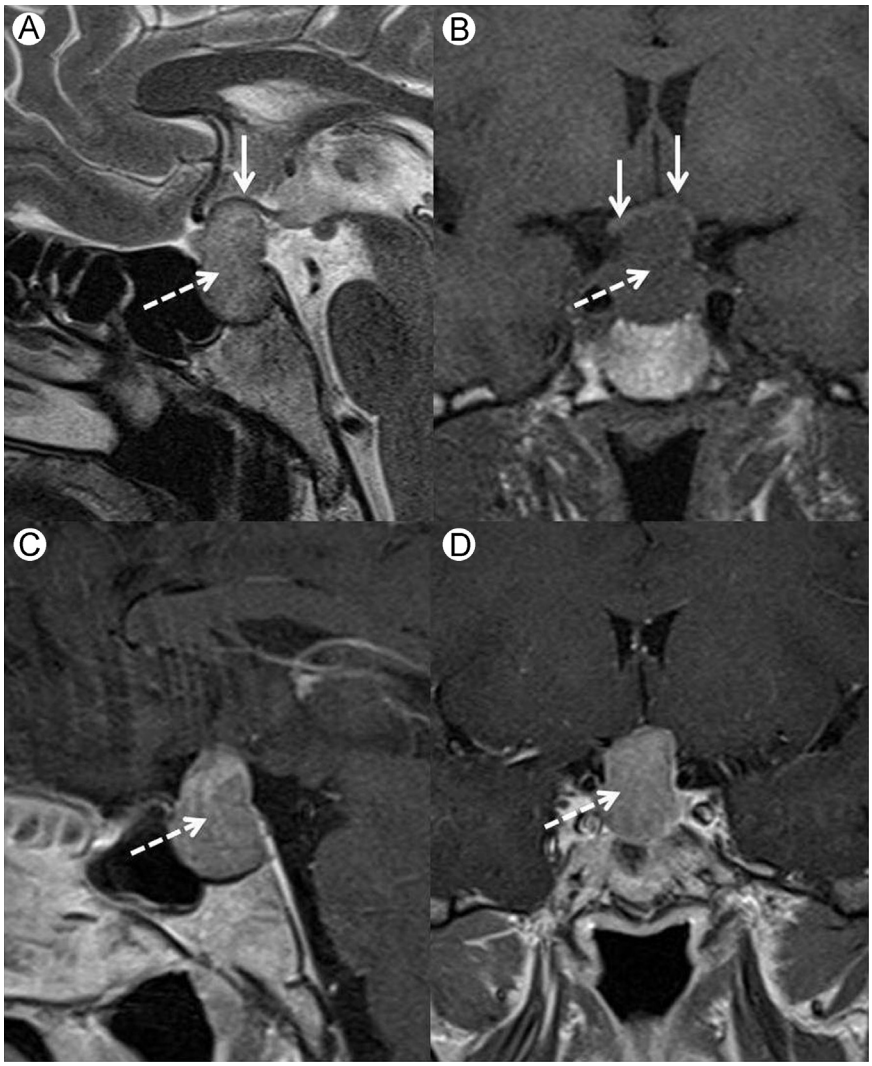


**Figure 4** A 32-year-old woman whose optician detected bitemporal hemianopia on routine examination: (A) sagittal T2-weighted; (B) coronal T1-weighted; (C) sagittal; and (D) coronal T1 postcontrast images of the pituitary gland showing a large "snowman-shaped" pituitary mass lesion (dashed arrow), compressing the optic chiasm (solid arrows, A and B), which is fixed posterior to the lesion.

Nevertheless, the anterior chiasmal syndrome is an established clinical entity,[37] and its identification, which may only be present on formal perimetry, allows absolute location of the lesion,[33] which in most cases is a pituitary adenoma.[38]

Usually, the body of the chiasm is compressed, resulting in a bitemporal hemianopia, although this may be asymmetrical and visual acuity preserved. The defect, although usually caused by a pituitary adenoma (Fig. 4), is indistinguishable from that caused by other suprasellar suprachiasmal lesions such as meningiomas and craniopharyngiomas. Papilledema may be a clinical sign that the lesion is suprachiasmatic and obstructing the third ventricle.[33]

Posterior chiasmal lesions produce bitemporal hemianopic scotomas owing to the interference with crossing macular fibers from both nasal hemiretinae. A degree of homonymous hemianopia may be present if the optic tract is involved. Visual acuity and color perception are usually spared—an important distinguishing feature from other causes of central scotomas, such as toxic and metabolic insults, where acuity and color vision are reduced.[33]

It is worth noting that infiltrating tumors such as gliomas (Fig. 5), inflammatory lesions, and demyelination around the optic chiasm do not tend to produce stereotypical visual field defects, and the visual loss may not correlate to the extent or location of the lesion.[33] The range of lesions affecting the chiasm makes MRI the preferred imaging modality.[25]

## Homonymous Visual Field Defects

Lesions posterior to the optic chiasm, that is, those of the optic tracts, LGN, optic radiations (ORs), or primary visual cortex, produce homonymous visual field defects without loss of acuity. Localization without additional clinical details (Fig. 6) is challenging, although generally neoplastic lesions produce a gradual onset of symptoms in contrast to the sudden onset associated with vascular lesions.[33]

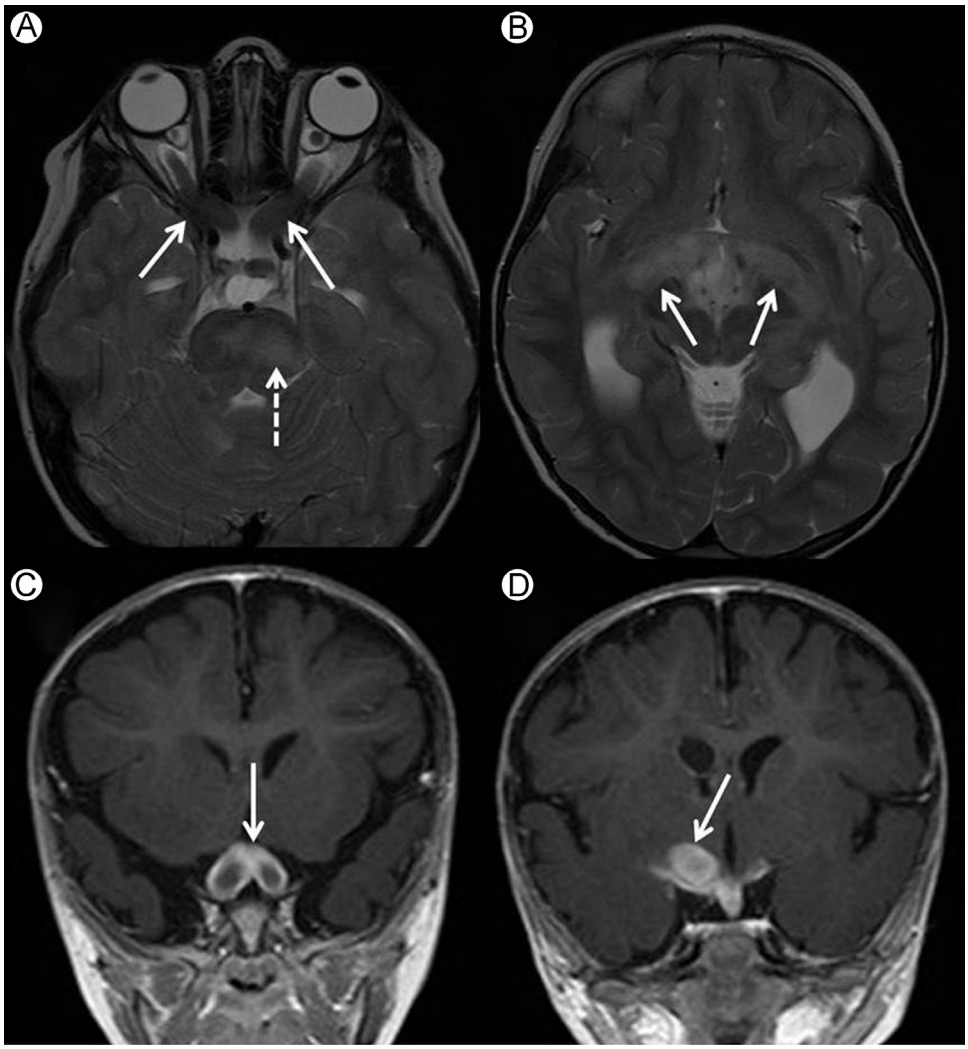


**Figure 5** A 4-year-old child with neurofibromatosis type 1. (A and B) Axial T2 and (C and D) coronal T1 postcontrast images. There is diffuse fusiform enlargement of the optic nerves (arrows, A) and T2 hyperintensity in the optic tracts (arrows, B) with avid peripheral enhancement of both the cisternal segments of the optic nerves (arrows, C) and right optic tract (arrow, D). An "NF-related" hyperintensity is seen in the left hemipons (dashed arrow, A).

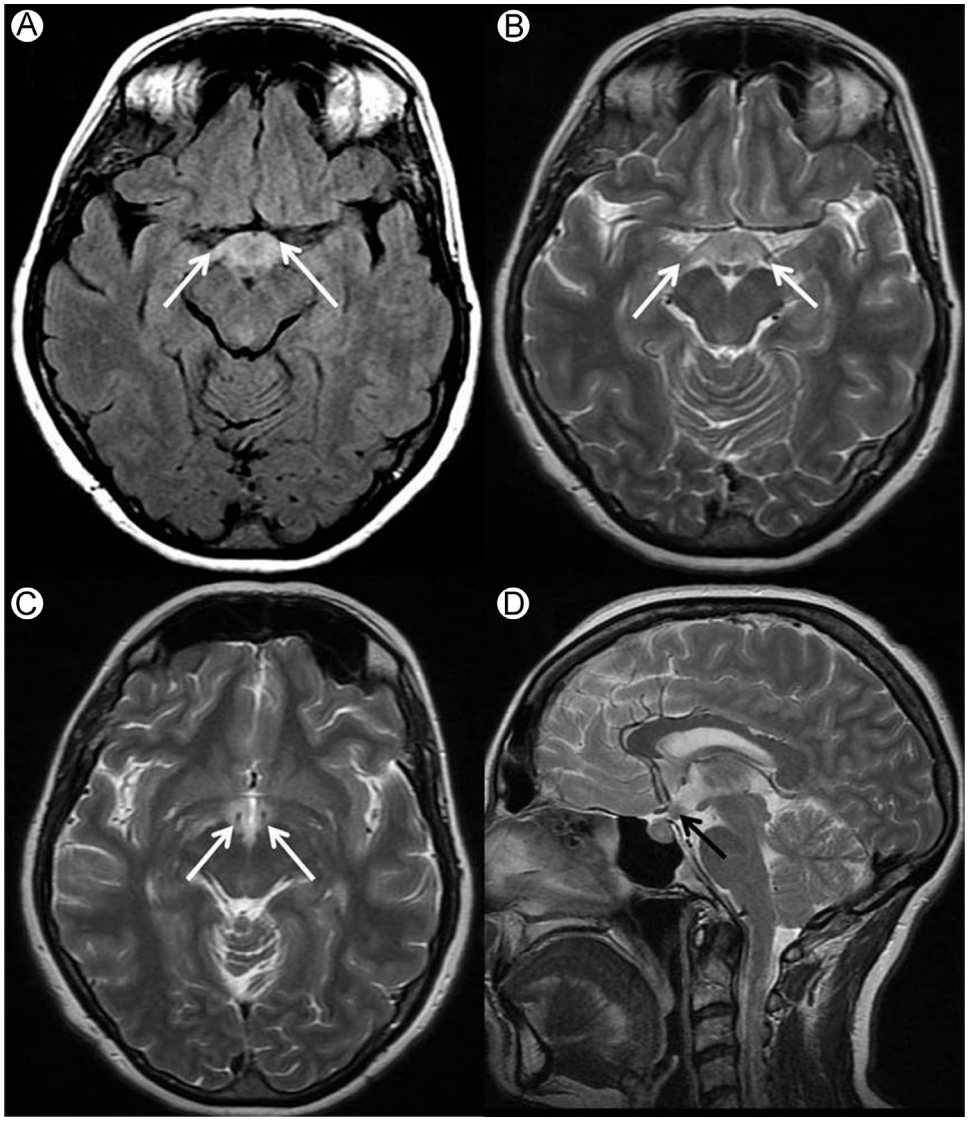


**Figure 6** A patient with metastatic breast cancer presenting with diabetes insipidus and visual impairment: (A) axial FLAIR; (B and C) axial T2-weighted; and (D) sagittal T2 images. There is edema in the optic tracts (arrows, A and B) and hypothalamic edema outlining the anterior columns of the fornix (arrows, C), due to a T2 hypointense metastasis in the anteroinferior third ventricle (arrow, D). FLAIR, fluid-attenuated inversion recovery.

## The Optic Tract

Homonymous hemianopias secondary to lesions of the optic tracts are rare and together with lesions of the LGN represent only 5%-11% of cases.[39,40] Clinically, an optic tract lesion should be suspected if there is homonymous hemianopia and a relative afferent pupillary defect contralateral to the side of the lesion with normal visual acuity and color vision.[33] The optic tracts are susceptible to lesions that affect the optic chiasm (Fig. 7) but can also be involved in pathology arising in the medial temporal lobes and midbrain (Fig. 8).[2] MRI is the modality of choice in demonstrating the variety of neoplastic, vascular, demyelinating, and inflammatory lesions, the characteristics of which are well reviewed elsewhere.[41] More recently, DTI has been used in the context of peritumoral edema to aid differentiation between pure vasogenic edema and tumor infiltration[42-45] with potential implications for surgical planning and treatment (Fig. 8).

## The Lateral Geniculate Nucleus

The lateral geniculate nucleus of the thalamus is located posterolateral to the pulvinar and is the main input to the visual cortex. The lateral geniculate nucleus is comprised of 6 layers of cell bodies numbered 1-6, ventral to dorsal. Spatial segregation is maintained, with the crossed or nasal retinal fibers projecting to layers 1, 4, and 6 and the uncrossed or temporal retinal fibers projecting to layers 2, 3, and 5. A functional division also occurs. Axons from the M-type retinal ganglion cells terminate in layers 1 and 2, the magnocellular (large cell) ventral layers. Axons from P-type retinal ganglion cells terminate in layers 3-6, the parvocellular (small cell) dorsal layers. M and P cells demonstrate

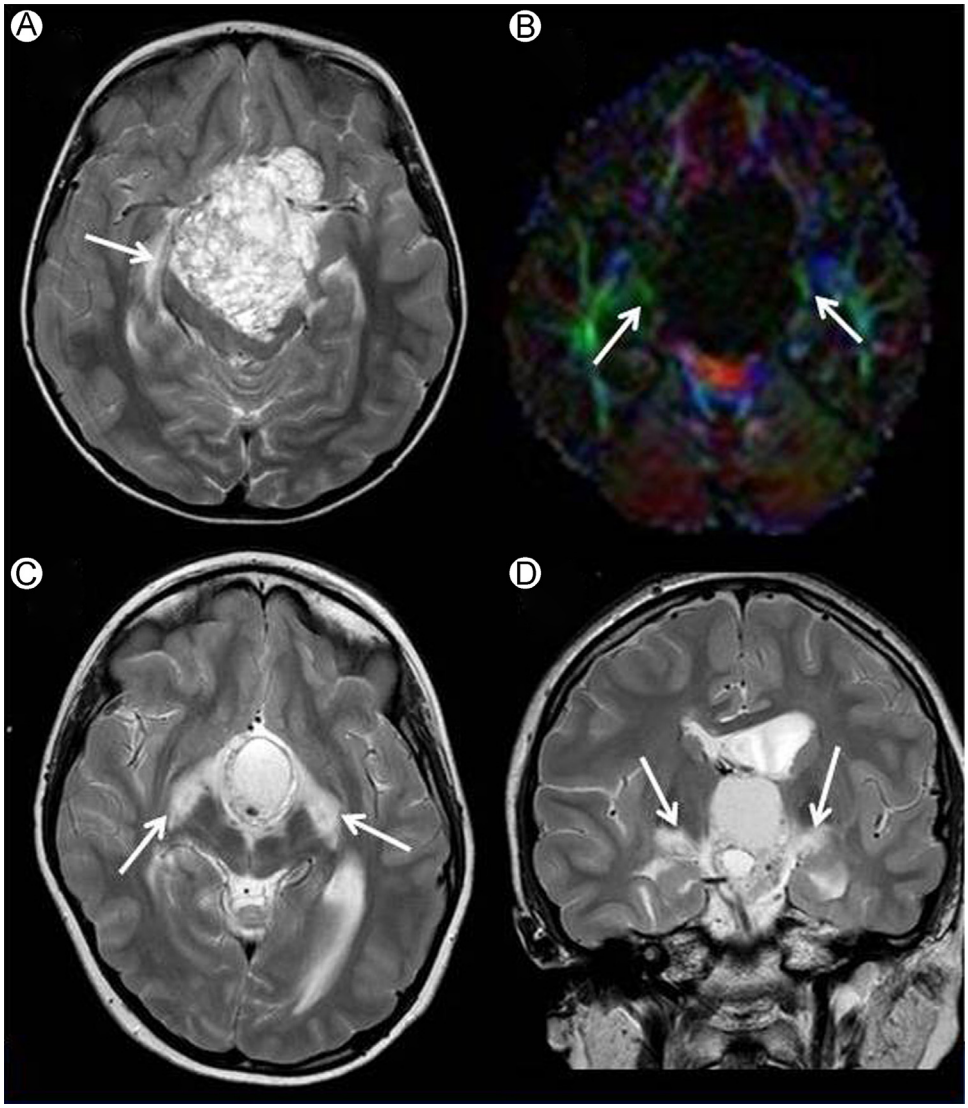


**Figure 7** Large cystic suprasellar craniopharyngioma. (A) Axial T2 and (B) color FA map. The optic tracts are displaced superolaterally (arrows, A and B) but the FA is maintained (arrows, B)—the patient did not have visual symptoms. (C and D) A different patient with a cystic craniopharyngioma. There is diffuse optic tract edema, despite only minor compression (arrows, C and D). This is a well-recognized feature seen frequently with this type of tumor. FA, fractional anisotropy.

contrasting properties, with M being more sensitive to lower contrast and higher temporal (ie, motion) frequencies than P cells are. Conversely, P cells are responsive to higher spatial frequencies and are notably sensitive to color contrast.[32,46,47] This functional division continues to the visual cortex and its significance is discussed later.

Lesions of the LGN are encountered less frequently than lesions of the optic tracts are and most frequently result from infarction of the anterior or lateral choroidal arteries, branches of the internal carotid and posterior cerebral arteries respectively.[2,48] The lateral and medial portions of the LGN represent the superior and inferior hemifields, respectively, and lesions involving the lateral or medial portions of the LGN cause a corresponding superior or inferior quadrantanopia (Fig. 9).[49,50] The anterior choroidal artery supplies both the medial and lateral portions, and distal disruption of this supply results in a quadruple sectoranopia, that is, an incomplete, usually peripheral, wedge-shaped homonymous hemianopia.[49,51] The lateral choroidal artery supplies the hilum of the LGN and occlusion results in a homonymous horizontal quadrantanopia.[50,52] Geniculate hemianopia was initially thought to be incongruous, that is, characterized by asymmetrical defects.[53] However, congruous defects, those that are similar in each eye, have been described, and it is suggested that partial lesions tend to cause incongruous defects, with congruent defects typically due to selective ischemia.[48,54]

## The Optic Radiation

Neurons from the LGN project through the retrolenticular portions of the internal capsules as the optic radiations (OR) or geniculocalcarine tracts. The inferior fibers contain information about the superior visual field and initially pass anteriorly as

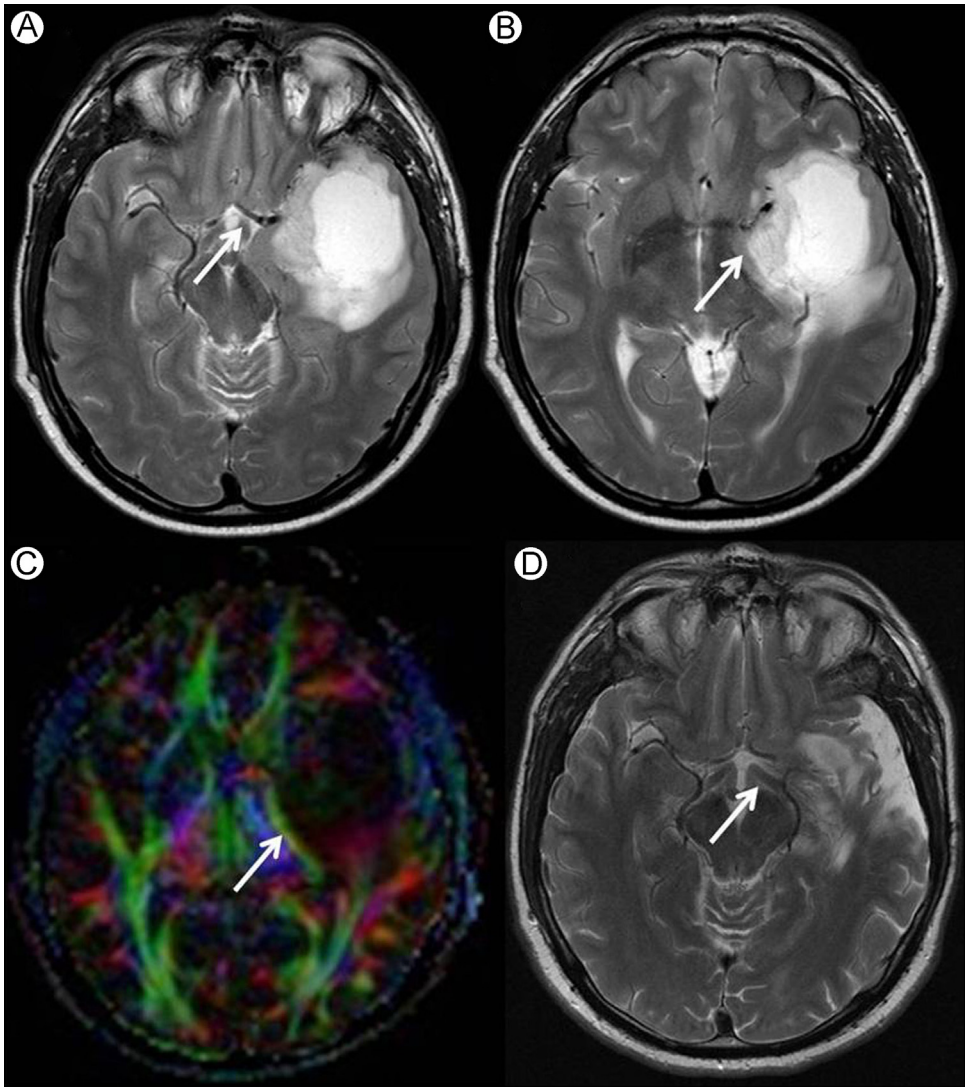


**Figure 8** A patient presenting with seizures and a right-sided homonymous hemianopia. (A, B, and D) Axial T2 and (C) color FA map. The left optic tract is displaced and hyperintense (arrows, A and B) but shows preserved visualization on color FA (arrow, C). Postoperative follow-up shows preservation and return to normal position of the left optic tract (arrow, D). Histology revealed a grade II oligodendroglioma. FA, fractional anisotropy.

Meyer loop, lateral to the anterior portion of the temporal horn of the lateral ventricle, then course through the temporal lobes to terminate in the primary visual cortex below the calcarine fissure in the medial surface of the occipital lobe. The superior tracts contain information regarding the inferior visual field and travel through the parietal lobe, form part of the wall of the superior aspect of the lateral ventricle, and terminate in the superior part of the primary visual cortex above the calcarine fissure.[30,31]

A contralateral superior quadrantanopia or wedge-shaped defect ("pie-in-the-sky" defect) results from lesions in the inferior temporal components of the OR and is more commonly neoplastic or infectious in origin rather than vascular.[33] Exact characterization of the anterior part of the OR into the temporal lobe, Meyer loop, is controversial and surgical, cadaveric and DTI tractography have generated differing and overlapping measurements of its anterior extent.[55] This is not solely of academic concern. Visual field defects following temporal lobe resection can be avoided or reduced with accurate characterization of the Meyer loop.[56] Tractography offers the only practicable method for preoperative assessment to reduce visual field defects but this currently requires further validation and standardization (Figs. 10 and 11).[55,57]

Lesions affecting the superior parietal projections of the OR cause contralateral inferior quadrantanopia (Fig. 11) or a predominantly lower homonymous field defect that tend to be more congruous (similar in each eye) than defects secondary to temporal lobe lesions.[33] Large lesions may produce a complete homonymous hemianopia with macular splitting as the entire OR passing through the temporoparietal lobe is affected. In these cases, infarction is the commonest cause, accounting for more than half of the cases.[40]

Lesions relating to the OR may be relatively clinically silent and only discovered as a result of investigation for visual field defects (Fig. 12). Conversely, lesions in the temporal or parietal

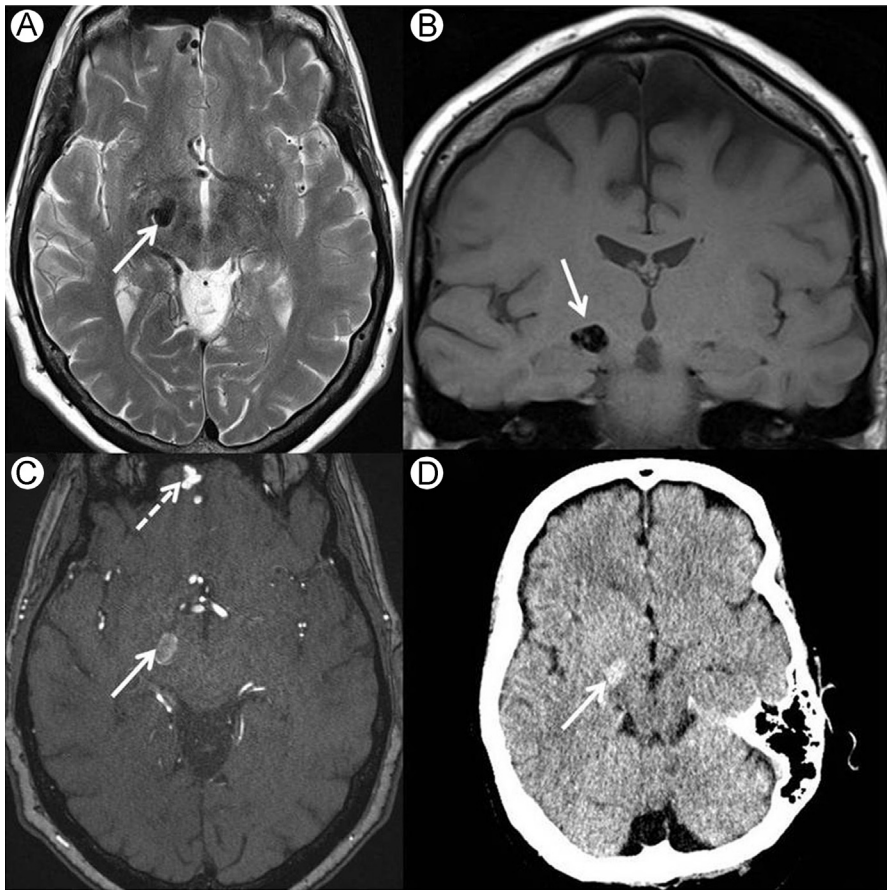


**Figure 9** A patient presenting with a left superior quadrantanopia: (A) axial T2-weighted, (B) coronal T1-weighted, (C) axial time-of-flight MR angiogram, and (D) unenhanced CT. There is a pial AV fistula supplied by the anterior ethmoid arteries (not shown). Dilated draining veins are seen in the anterior hemispheric fissure (dashed arrow, C), a dilated venous pouch (arrows, A-D) is compressing the right lateral geniculate nucleus. AV, arteriovenous; CT, computed tomography.

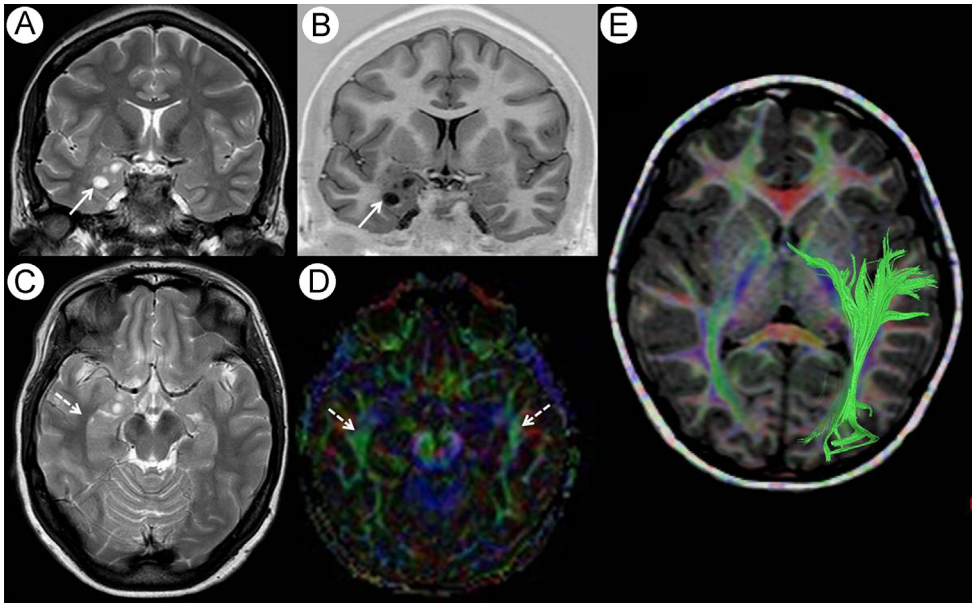


**Figure 10** Right medial temporal lobe dysembryoplastic neuroepithelial tumor (DNET, arrows A and B) in a patient with temporal lobe epilepsy. (A) Axial and (C) coronal T2, (B) coronal T1-inversion recovery, (D) color FA map, (E) left optic radiations tractography of a normal patient, superimposed on fused color FA map-T1-gradient echo volume. The location of the right Meyer loop can be estimated on T2 (arrow, C) and confirmed on color FA map (dashed arrows, D). The proximity of the Meyer loop to the tumor should be appreciated at the preoperative stage. FA, fractional anisotropy.

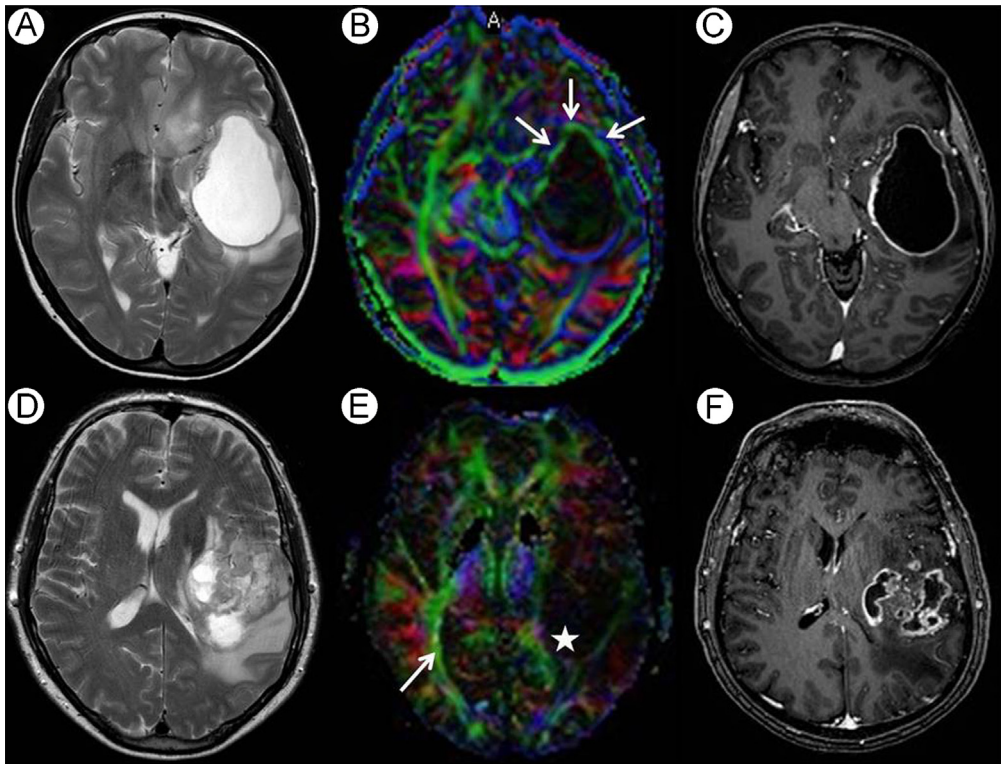


**Figure 11** Two patients with glioblastoma multiforme (GBM). (A and D) Axial T2, (B and E) color FA map, and (C and D) postcontrast T1-gradient echo volume. (A-C) A large cystic temporal lobe GBM displaces the Meyer loop anteriorly (arrows, B). In the bottom row, a patient with a multicystic necrotic temporoparietal tumor shows destruction of the parietal fibers of the left optic radiation (compare normal side—arrow to the destroyed side—star, E). The top patient did not suffer from a visual field defect after resection whereas a left inferior quadrantanopia present in the second patient persisted after resection. FA, fractional anisotropy.

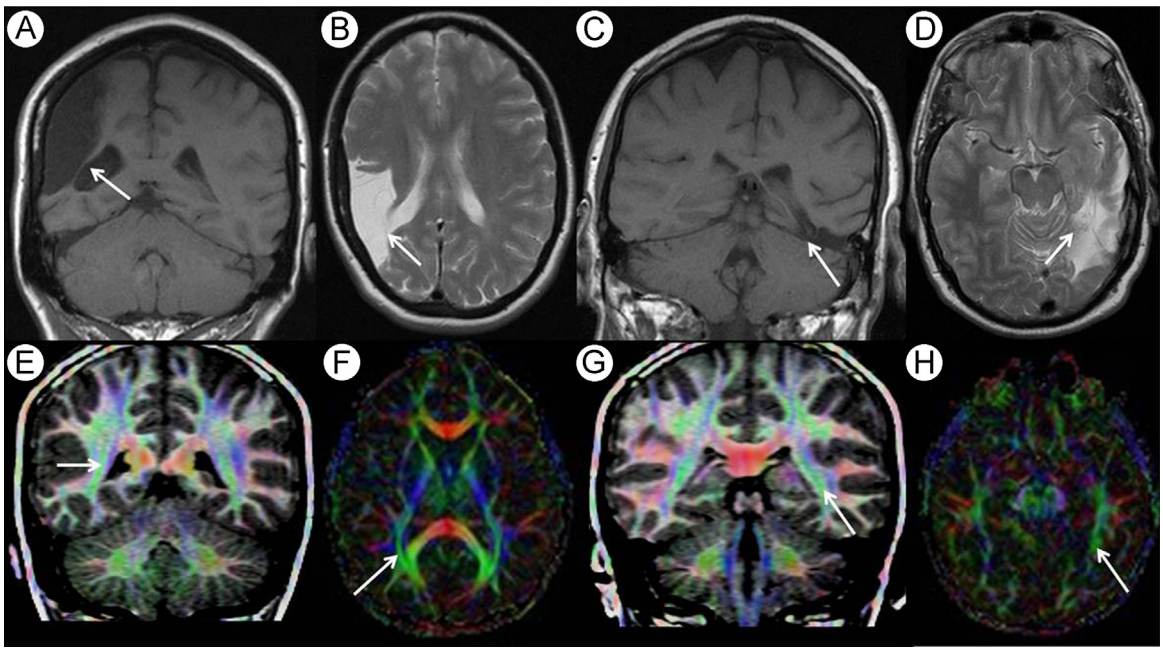


**Figure 12** Quadrantanopia in 2 patients. (A) Coronal T1- and (B) axial T2-weighted images in a patient presenting solely with a left inferior quadrantanopia. There is a large, mature right inferior divisional MCA infarct (arrows, A and B). (C) Coronal T1- and (D) axial T2-weighted images in a second patient presenting with a right superior quadrantanopia show a left inferior temporal infarct (arrows, C and D). Fused color FA and T1 volumes (E and G) and axial color FA maps (F and H) showing the correlating normal positions of the optic radiations affected in the cases above. FA, fractional anisotropy; MCA, middle cerebral artery.

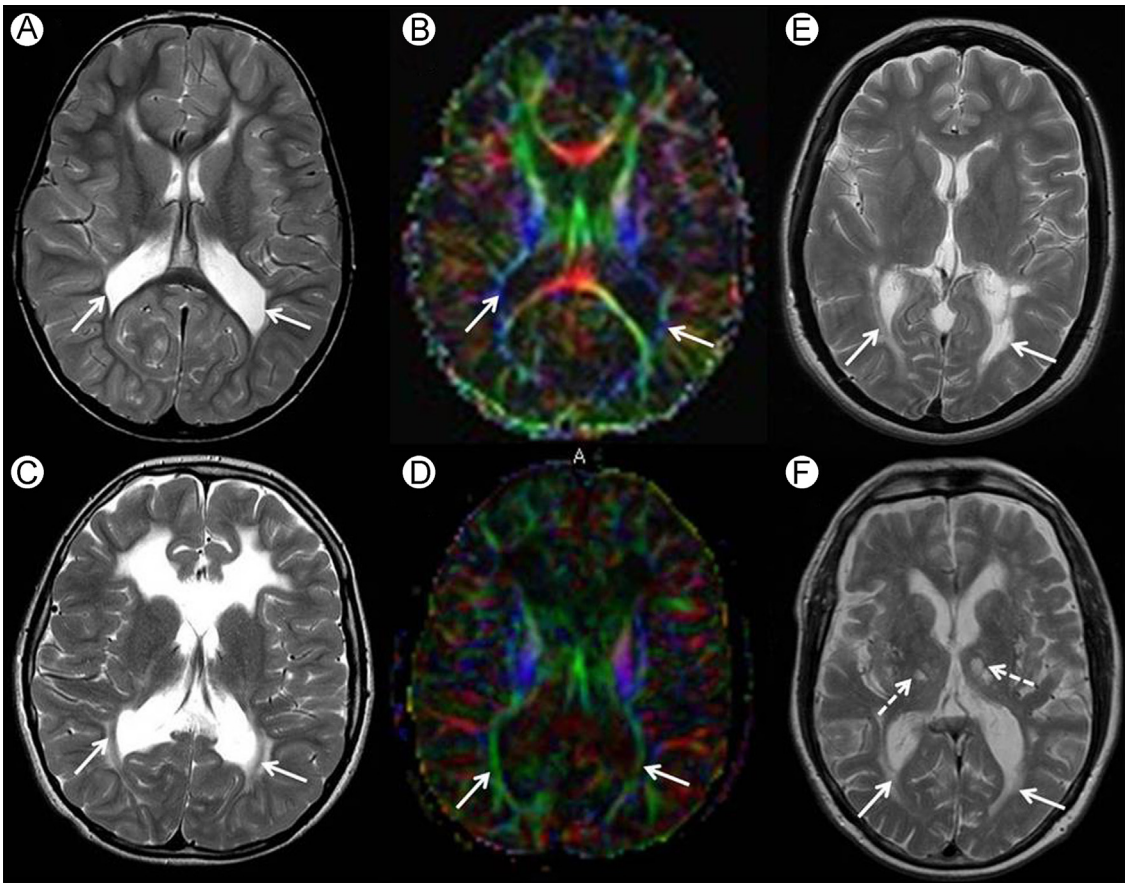


**Figure 13** White matter disease affecting the optic radiations. (A) Axial T2 and (B) color FA map in a child with extensive PVL. There is severe loss of periventricular white matter and corresponding loss of visualization of the optic radiations on DTI (arrows, A and B). (C) Axial T2 and (D) color FA map in a second child with frontal variant of adrenoleukodystrophy shows relative preservation of optic radiations (arrows, C and D) compared with extensive destruction and lack of visualization of normal frontal white matter (images A-D courtesy of Dr T Jaspan, NUH, UK). (E) Axial T2-weighted image of a patient with multiple sclerosis with diffuse periventricular disease affecting the optic radiations (arrows). (F) Axial T2-weighted image of an elderly patient with extensive small vessel ischemic change shows T2 hyperintensity involving the optic radiations (arrows) and mature lacunar infarcts in the thalami (dashed arrows). FA, fractional anisotropy; PVL, periventricular leukomalacia.

lobes may present with nonvisual symptoms.[40] These may cause seizures and auditory, vestibular, speech, and memory disturbances when involving the temporal lobe and paresthesia, inattention, neglect, apraxias, agnosias, and speech disturbances when involving the parietal lobe.[58] Diffuse white matter lesions such as periventricular leukomalacia adrenoleukodystrophy in children or infants and MS and small vessel ischemic disease in adults may affect both ORs and cause mixed, nonspecific visual field defects (Fig. 13).

## The Primary Visual Cortex

The primary visual or striate cortex is located on the medial surfaces of the occipital lobes above and below the calcarine fissures. As with the preceding components of the visual pathway, an anatomical map of the visual field is preserved. The most caudal part of the primary visual cortex, extending to the occipital poles, represents the fovea and the volume of tissue is relatively large compared with the area of the retina that the fovea occupies. More rostral portions of the cortex represent increasingly peripheral regions of the visual field. The cortex above the calcarine fissure represents the inferior visual field and vice versa, with each hemisphere representing the contralateral visual field.[30-32] Of the 6 cortical layers, the major input layer from the lateral geniculate nucleus is layer 4, which is prominent in the primary visual cortex. Functional segregation between motion and visual content information is preserved, with M and P axons terminating principally in sublayers 4Cα and 4Cβ.[32]

Lesions affecting the occipital lobe account for nearly half of homonymous field defects with infarction, secondary to middle cerebral and posterior cerebral arterial supply, accounting for nearly 75% of these lesions (Figs. 14 and 15).[40] Lesions of the occipital pole produce contralateral homonymous scotomas that are extremely congruous. More anterior lesions progressively involve the peripheral vision and again are highly congruous a feature that can be used to differentiate such lesions from those of the ORs or optic tracts (Fig. 14).[33] Lesions at the very anterior edge of the primary visual cortex may

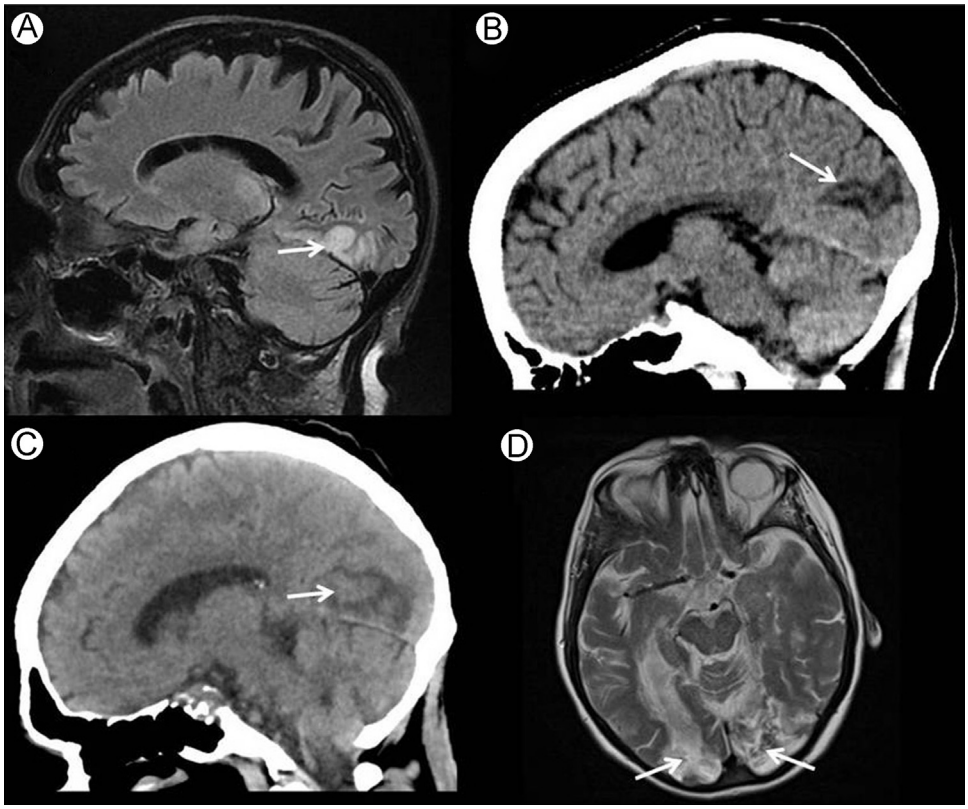


**Figure 14** Occipital infarcts in 4 different patients: (A) sagittal FLAIR image showing inferior lip calcarine cortical infarct (arrow) in patient with superior quadrantanopia; (B) sagittal CT in a patient with an inferior quadrantanopia due to a superior lip calcarine infarct (arrow); (C) sagittal CT in a patient with homonymous hemianopia due to involvement of both superior and inferior lips of the calcarine cortex (arrow); and (D) axial T2-weighted image of a patient with cortical blindness due to bilateral infarcts of the occipital poles (arrows). CT, computed tomography; FLAIR, fluid-attenuated inversion recovery.

produce peripheral monocular temporal defects, a temporal crescent. This defect is monocular because the relatively larger temporal visual field has no nasal counterpart in the post-chiasmatic visual pathway. Although this locates the lesion to the anterior striate cortex, it is very rarely encountered[40] and most causes of a peripheral monocular temporal field defect are retinal.[33] Lesions above and below the calcarine fissure produce inferior and superior quadrantanopias, respectively (Fig. 14). Classically, a homonymous hemianopia with macular sparing has been cited as useful in locating a lesion to the occipital lobe, although a recent large case series found that homonymous hemianopia with macular sparing is due to lesions without the occipital lobes in nearly half of the cases.[40]

Bilateral occipital lobe lesions occurring either simultaneously or, more usually, sequentially can produce any combination of bilateral homonymous hemianopia with or without macular sparing and of varying degrees of congruity.[33] When damage to the primary visual cortex is complete, cortical blindness results and the lesion usually extends beyond the primary visual cortex. Interesting observations have been made in some patients with cortical blindness, which include "blindsight," the ability to respond to visual stimuli without awareness and Anton syndrome, the denial of cortical blindness.[59] Some attempts have been made to explain the neural mechanisms underlying blindsight using tractography and implicate connections from the LGN and superior colliculi to higher visual centers bypassing the primary visual cortex.[60,61]

## Disorders of Visual Perception

Overall, 2 pathways emerge from the primary visual cortex: a dorsal pathway extending into the parietal lobe and a ventral pathway extending to the temporal lobe. Although not exclusive, there is some degree of preservation in the division of the M and P fibers from the LGN to the dorsal and ventral pathways, respectively.[62] Accordingly, perception of motion appears to occur primarily in the dorsal or parietal pathway and perception of object form and color in the ventral or temporal pathway. These have been termed the "where" and "what" pathways, respectively, although these "streams" have no clear anatomical boundaries.[63]

## The Ventral Stream

Cerebral achromatopsia results from lesions in the ventromedial aspect of the occipital lobe and patients report seeing in shades of gray or feel their perception is less bright. The finding is rarely isolated and occurs in a

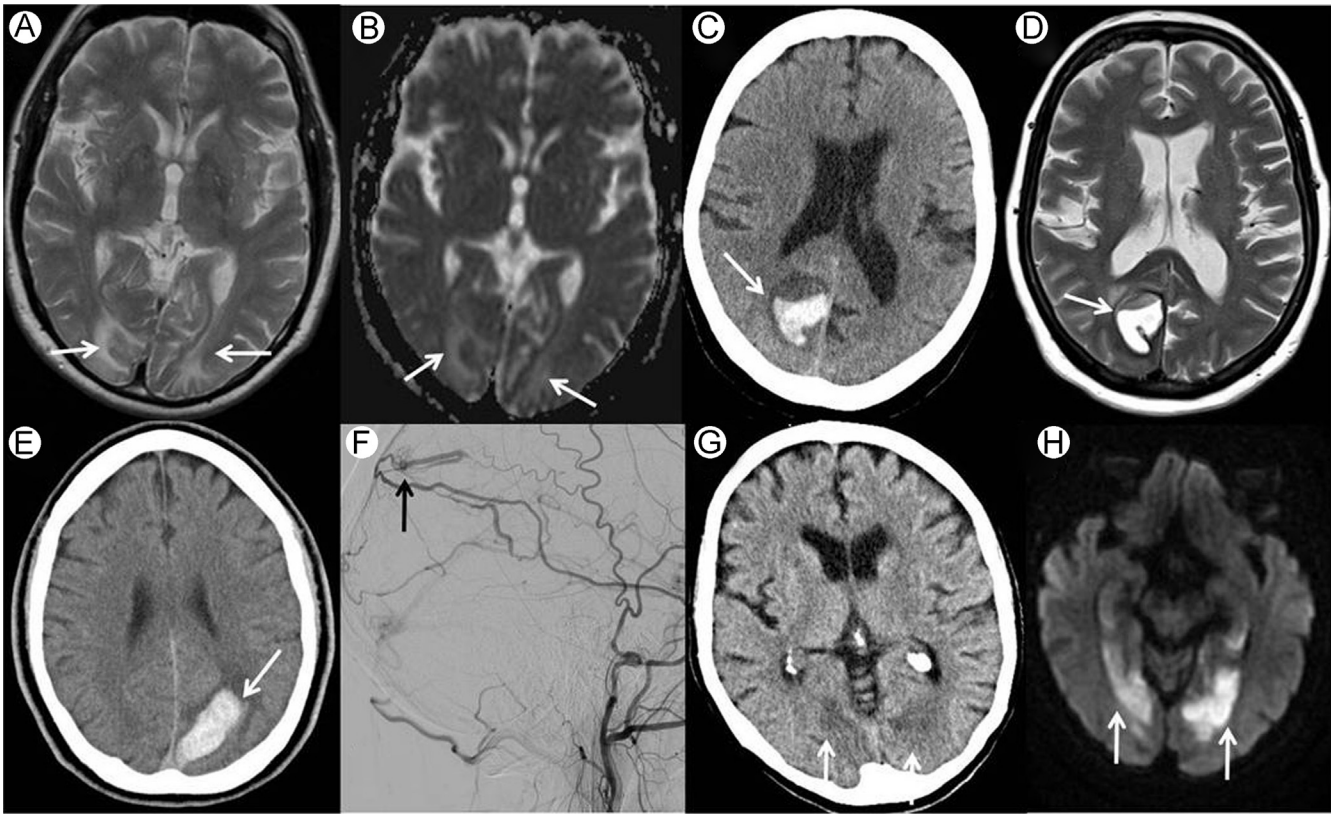


**Figure 15** Occipital lobe lesions. (A) Axial T2 and (B) ADC map in a patient with posterior reversible encephalopathy syndrome (PRES) showing bilateral occipital subcortical white matter T2 hyperintensity and increased diffusivity (arrows). (C) Axial CT and axial T2-weighted image of a hemorrhagic right occipital infarct (arrows, C and D). Note that, although swollen, the boundary of the PCA territory is respected. (E) Axial CT of a 34-year-old patient with left occipital hemorrhage secondary to a dural AV fistula (arrow). Note the lesion crosses the PCA territory boundary. (F) External carotid artery injection catheter angiogram in the same patient demonstrating middle meningeal supply of the fistula (arrow). (G) Axial CT and (H) DWI demonstrating bilateral posteromedial temporo-occipital infarcts (low attenuation on CT, arrows, G and showing diffusion restriction, arrows, H) in a patient presenting with prosopagnosia due to bilateral infarction of the fusiform face area. ADC, apparent diffusion coefficient; AV, arteriovenous; CT, computed tomography; DWI, diffusion-weighted imaging.

tetrad with prosopagnosia (described later), a superior quadrantanopia and topographagnosia (agnosia for landmarks, resulting in getting lost in familiar locations).[59] Modern imaging studies suggest lesions affecting lingual and fusiform gyri on the ventromedial aspect of the occipital lobes, areas V4 and V8, are instrumental in causing cerebral achromatopsia.[64,65]

Prosopagnosia, the inability to recognize familiar faces, classically results from bilateral lesions to the lingual and fusiform gyri most commonly secondary to posterior cerebral artery infarction and head trauma (Fig. 15).[59] A single region does not appear solely responsible for facial recognition in humans[66,67] and experimental data confirm 3 important locations: the fusiform face area located on lateral aspect of the midfusiform gyrus; a face-selective region in the posterior superior temporal sulcus; and the occipital face area, posteriorly in the inferior occipital cortex.[68,69] A lesion study demonstrated that lesions most frequently involve the occipital facial area.[64]

Acquired alexia, the inability to read in previously literate subjects with normal visual acuity, usually occurs secondary to infarction in the left posterior cerebral artery territory, resulting in damage to the inferior occipitotemporal region.[59,70] Lesion overlap studies demonstrate medial occipitotemporal lesions extending to the lateral occipitotemporal junction.[70] The visual word form area, located in the left lateral occipitotemporal sulcus, and its connection to the occipital cortex via the inferior longitudinal fasciculus (ILF) appear crucial in reading with alexia resulting from lesions to the visual word form area and ILF (Fig. 16).[71,72]

## The Dorsal Stream

Balint syndrome comprises a triad of deficits originally described in a patient with bilateral parietal lobe lesions. The syndrome consists of the inability to comprehend the totality of a picture or scene, simultanagnosia; the impairment of visually guided grasping or reaching, despite adequate strength and co-ordination, optic ataxia; and the inability to shift gaze voluntarily, optic apraxia.[73,74] Balint syndrome has also been described in bifrontal lesions and simultanagnosia with superior occipital lobe, parieto-occipital fissure, and right

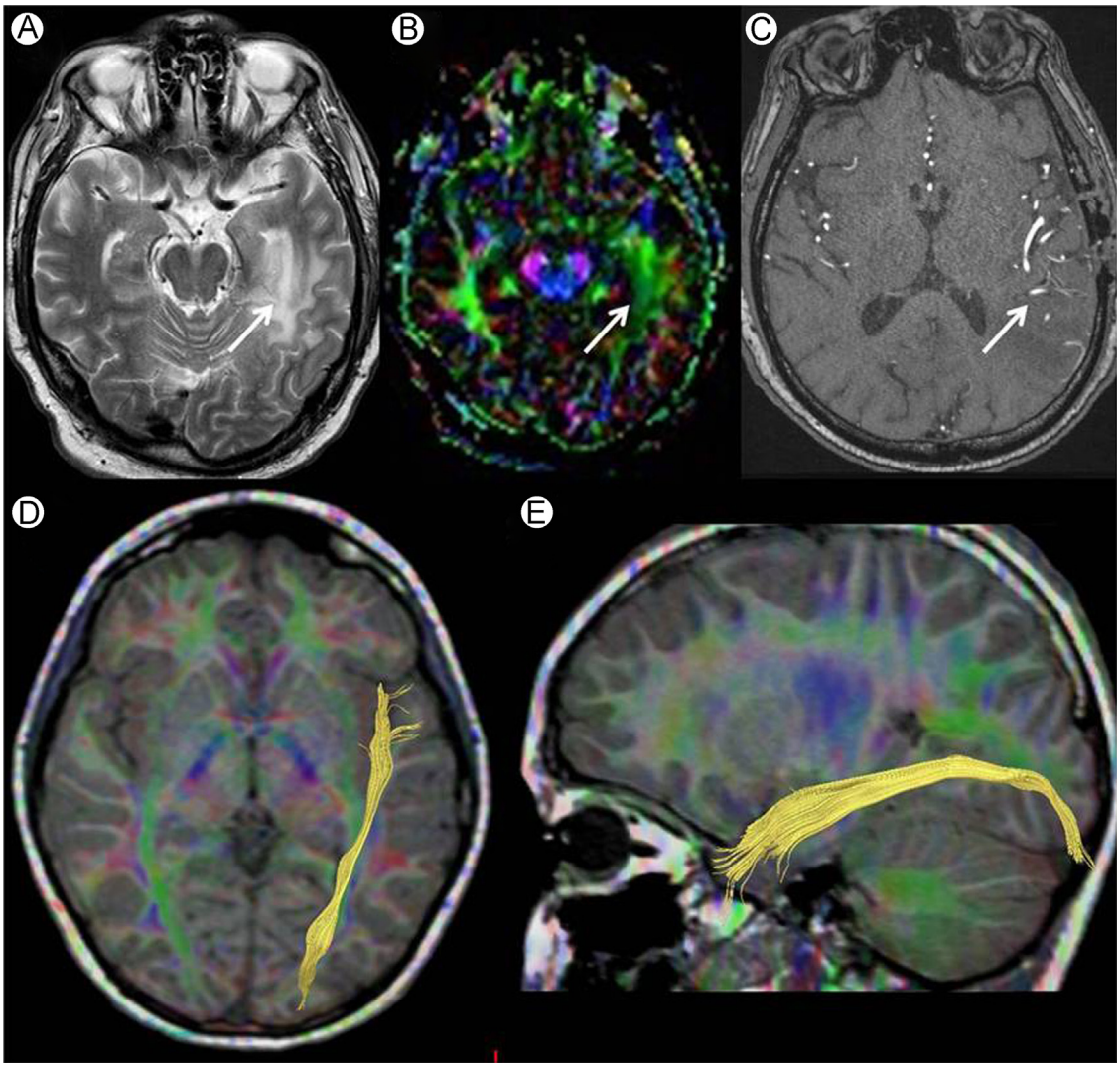


**Figure 16** A patient presenting with an inability to read his newspaper. (A) Axial T2, (B) color FA map, and (C) time-of-flight MR angiogram in a patient with biopsy-proven left temporal cerebral vasculitis. There is T2 hyperintensity in the left inferior temporal white matter (arrow, A) and reduced FA in the left inferior temporo-occipital fasciculus (arrow, B). Increased vascular engorgement is demonstrated on MRA (arrow, C). (D and E) Tractography of a normal left inferior temporo-occipital fasciculus superimposed on fused color FA/T1 volume for comparison. FA, fractional anisotropy; MRA, MR angiography.

hemisphere parietal lesions.[59,75] Tractography has revealed associations with white matter pathways within the higher visual pathways, including the superior longitudinal fasciculus, the inferior fronto-occipital fasciculus, and the ILF (Fig. 17).[75,76]

## Disorders of Visual Gaze

A proportion of retinal ganglion cells project directly to the superior colliculi of the tectum in the dorsal aspect of the midbrain. Each superior colliculus sends projections to the pulvinar nucleus of the thalamus and then to the cerebral cortex, as well as receiving striate and extrastriate cortical inputs. The superior colliculi play a key role in the control of saccades—rapid gaze-shifting eye movements—which are initiated in the cerebral cortex.[32] The neural networks involved in gaze control in humans have been corroborated with tractography and confirm the pulvinar is interconnected with the superior colliculus, thalamus, caudate nucleus, and cortical targets in the primary and secondary visual areas, visual inferotemporal areas, posterior parietal association areas, and the frontal eye fields.[77] Functional imaging and tractography have established the frontal eye fields in the dorsolateral frontal lobe, the supplementary eye fields in dorsomedial frontal lobe, and the parietal eye fields as essential in voluntary eye movement and that there is significant interconnectivity (within and across hemispheres) and right hemisphere dominance.[78] Lesions in these areas or their interconnections can result in disordered voluntary gaze (refer to section on Balint syndrome). Forward gaze is a functional balance of influence of both frontal eye fields; when one side is damaged, such as by a large frontal infarct, the normal hemisphere is unopposed and "pushes" the eyes toward the side of the lesion. This appearance is known in the radiology literature as Prévost sign (Fig. 18).[79]

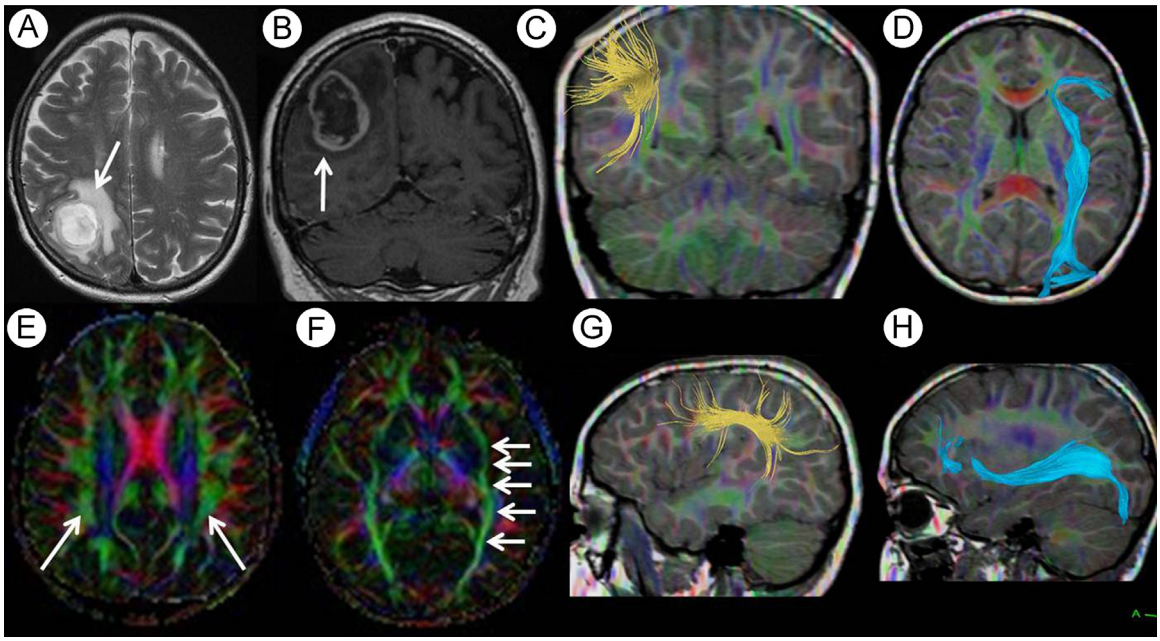


**Figure 17** A patient presenting with an inability to pick objects up when placed in front of her. (A) T2 axial and (B) T1 coronal postcontrast images of a right parietal glioblastoma (arrows). Examination revealed right hemifield visual neglect and simultagnosia. There is potential involvement of the SLF (normal comparison tractography, C and G; arrows on color FA map, E) or IFOF (normal comparison tractography, D and H; arrows on color FA map, F) or both. FA, fractional anisotropy; IFOF, Inferior fronto-occipital fasciculus; SLF, superior longitudinal fasciculus.

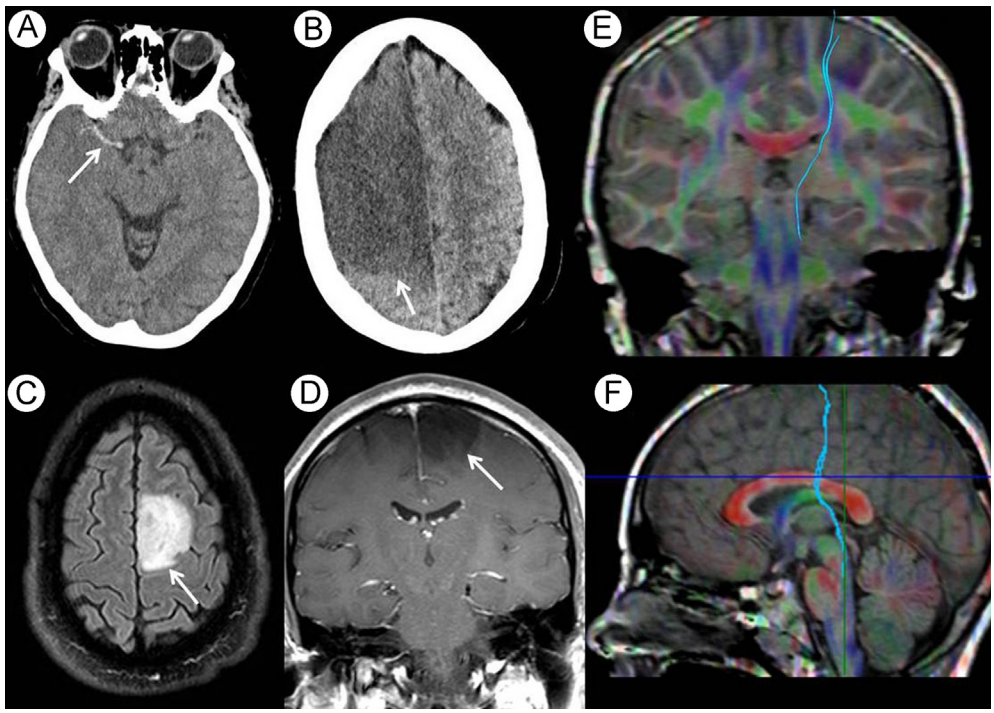


**Figure 18** Frontal eye fields. (A and B) Axial CT in a patient with right distal ICA occlusion and total right anterior circulatory infarction. There is a hyperdense right MCA (arrow, A), the eyes are deviated toward the side of the abnormality and there is extensive superior frontal infarction including the right frontal eye fields (arrow, B). (C) Axial FLAIR and (D) coronal T1 postcontrast images of a left superior frontal grade II astrocytoma (arrows, C and D). The patient presented with seizures with fixed rightward gaze due to hyperactivity in the contralateral frontal eye field. (E and F) Tractography acquired by placing regions of interest in the frontal eye fields and tectal plate (superimposed on fused color FA map-T1 volume) showing fibers passing from the frontal eye fields, through the pulvinar, to the tectal region. CT, computed tomography; FA, fractional anisotropy; FLAIR, fluid-attenuated inversion recovery; ICA, internal carotid artery; MCA, middle cerebral artery.

Eye deviation is often part of the onset of seizures and may be helpful in hemispheric localization. In this situation, increased electrical activity in one frontal eye field acts to "push" the eyes away from the side of the lesion.

## Conclusion

Intracranial lesions causing visual defects are many and varied. However, the functional deficit created by even small lesions can be clinically significant. A thorough understanding of the neuroanatomy serving visual function outlined previously can prompt a dedicated search strategy in such patients, aided by visualization of white matter tracts by DTI.

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