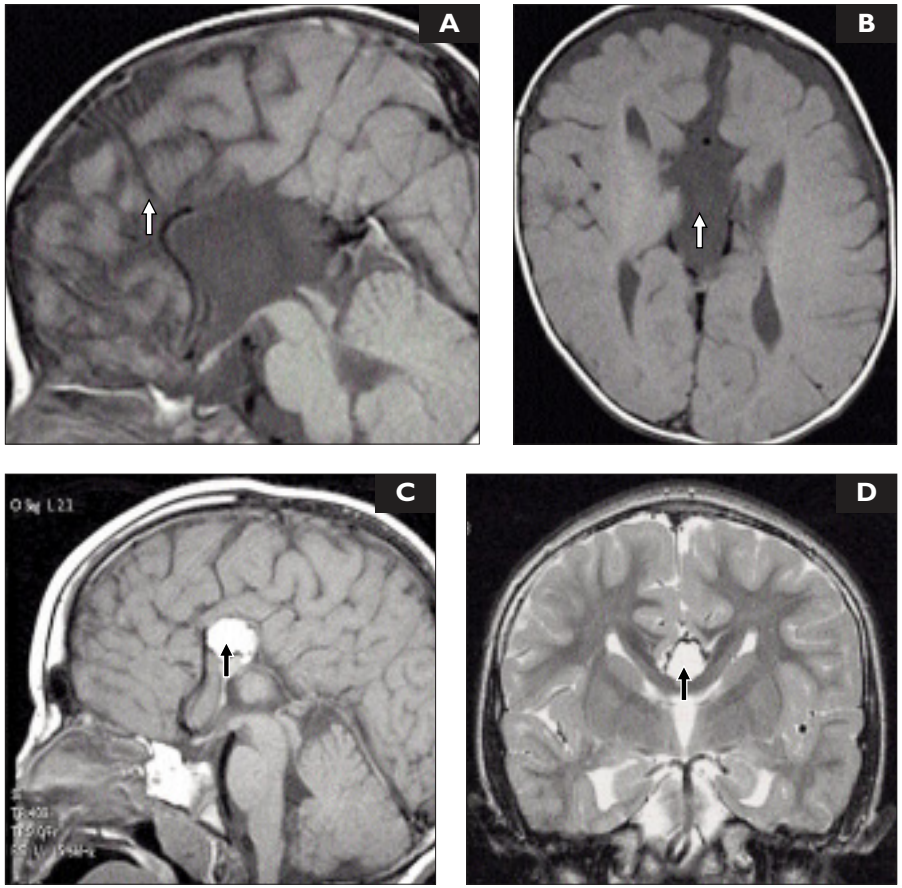
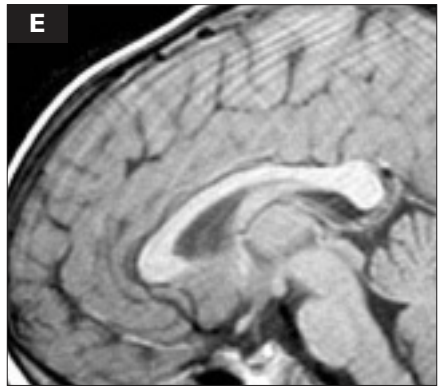


1 A 6-month old child presents with seizures.
i. Describe the features shown on imaging (**1A, B**).
ii. What is the diagnosis?
iii. What are its associations?
iv. How does the companion case shown (**1C, D**) differ?

1 Diagnosis
Agenesis of the corpus callosum (**ii**).



Imaging findings
The sagittal T1-weighted MRI image (**1A**) reveals agenesis of the corpus callosum (ACC). This is largest white matter commissure in the brain, lying in the midline, where it connects the two cerebral hemispheres (**1E**, arrow 1). Normal development of the corpus callosum is associated with inversion of the cingulate gyrus forming the cingulate sulcus, which parallels the corpus callosum superiorly (**1E**, arrow 2). Thus, in ACC, the cingulate sulcus is absent, causing the medial hemispheric sulci to radiate into the roof of the 3rd ventricle (**1A**, arrow). The axial T1-weighted MRI (**1B**), reveals the resultant high-riding 3rd ventricle extending superiorly between parallel lateral ventricles (arrow). The trigones and occipital horns of the lateral ventricles are often dilated in ACC, termed colpocephaly (**i**).

Pathology and clinical correlation
The corpus callosum forms during the first trimester and its absence (agenesis) or incomplete formation (hypogenesis) may result from one of many different genetic mutations. Associated developmental anomalies are common and include Chiari II malformation, Dandy–Walker malformation, and disorders of neuronal migration and organization. ACC is also a feature of many different syndromes, for example Aicardi syndrome. In this X-linked syndrome, ACC is associated with an interhemispheric cyst, cortical dysplasia (commonly polymicrogyria), grey matter heterotopia, and hypoplasic cerebellum (**iii**).

COMPANION CASE

Diagnosis
Hypogenesis of the corpus callosum with interhemispheric lipoma.

Imaging findings
The sagittal T1-weighted MRI image (**1C**) shows that only part of the corpus callosum has formed (anterior body). The posterior body, splenium, and rostrum of the corpus callosum are absent. On both the T1-weighted image and the coronal T2-weighted image (**1D**), a focal hyperintense mass is seen in the midline, posterior and superior to the formed corpus callosum (arrow). This is an interhemispheric lipoma, which is almost always associated with agenesis or hypogenesis of the corpus callosum.

Pathology and clinical correlation
The corpus callosum develops in an orderly fashion starting with posterior genu, body, anterior genu, splenium, and lastly rostrum. Knowledge of this normal pattern of development makes it is possible to differentiate a hypogenetic corpus callosum from one that has been secondarily damaged. Interhemispheric lipoma develops from abnormal differentiation of the mesenchyme that surrounds the developing brain (meninx primitiva).

Teaching pearls
➢ *Agenesis of the corpus callosum is commonly associated with other intracranial developmental anomalies.*

Reference
Barkovich AJ, Norman D (1988). Anomalies of the corpus callosum: correlation with further anomalies of the brain. *AJNR* 9:493–501.

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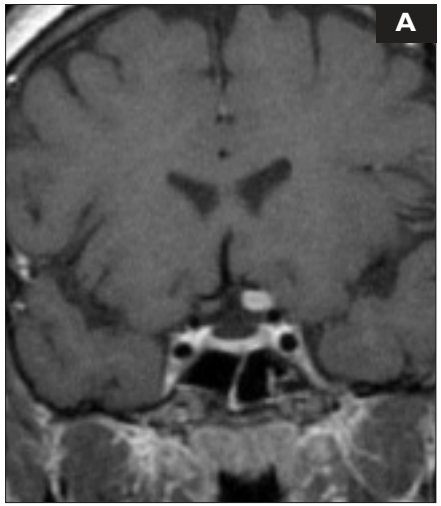
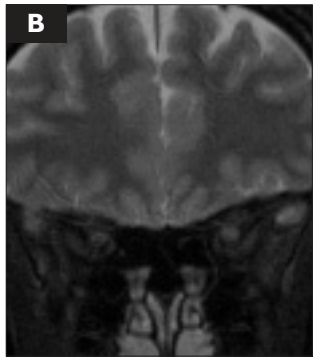
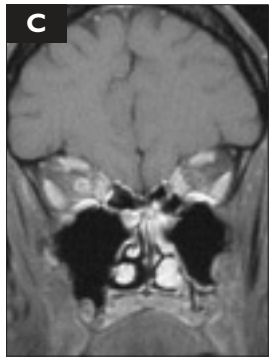
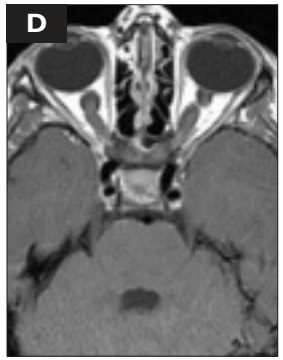
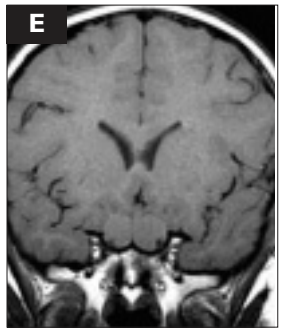
2 A 20-year-old female patient presents with decreased vision in her left eye.
i. Define the abnormal structure in image **2A**.
ii. Is extraocular muscle enhancement normal?
iii. Is the optic nerve encased by Schwann cells or meninges?











2 Diagnosis
Optic neuritis.

Imaging findings
Coronal T2 image (**2B**) shows subtle diffuse increased signal throughout the substance of the left optic nerve. Postcontrast T1 image (**2A**) reveals corresponding diffuse enhancement and confirms enlargement of the nerve (**i**). No perineural mass is present. The right optic nerve appears normal. Note that the extraocular muscles normally enhance (**ii**).

Companion cases (different patients) demonstrate perineural enhancement on postcontrast T1 coronal image in **2C** in a patient with an optic nerve meningioma. Images **2D** and **2E** are nice examples of severe bilateral nodular optic nerve enlargement due to bilateral optic nerve gliomas.

Differential diagnosis
The primary consideration when enhancement is noted related to the optic nerve is differentiation between enhancement of the actual nerve and enhancement of the adjacent meningeal lining (optic nerves are lined by meninges rather than schwann cells seen with peripheral nerves [**iii**]). In this case (**2A, B**), the nerve itself is clearly involved.

When nerve enhancement is present, the next determination is whether or not there is mass-like enlargement to suggest optic glioma. This case demonstrates only minimal diffuse enlargement and associated oedema (T2 hyperintensity). The primary differential considerations are optic neuritis or optic nerve vasculitis (most commonly related to infection, radiation, or autoimmune disorders). Optic neuritis is much more common and is the correct diagnosis in this case.

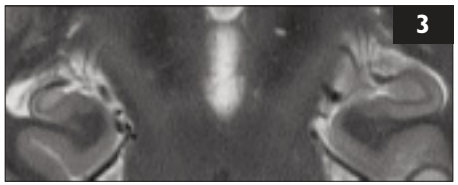
Pathology and clinical correlation
Though postcontrast fat saturation sequences show a 95% sensitivity in identifying optic nerve enhancement, MRI is not necessary for diagnosis of optic neuritis in the majority of cases. Occasionally, patients with nonarteritic anterior ischaemic optic neuropathy, acute compressive neuropathy secondary to a pituitary tumour or aneurysm, or posterior scleritis have symptoms mimicking optic neuritis and MRI provides the correct diagnosis. More commonly, the diagnosis of optic neuritis is known and the clinical question is whether or not the neuritis is an isolated finding. There is a strong association between optic neuritis and multiple sclerosis (MS). Indeed, approximately 20% of MS patients initially present with optic neuritis. Furthermore, 50–60% of patients with optic neuritis will be diagnosed with MS over the subsequent 15 years.

Interestingly, the length of nerve enhancement with optic neuritis appears to correlate with initial severity of visual impairment (both colour perception and special resolution). However, there is no correlation between degree of enhancement and final recovery of function. Steroids are administered as the mainstay of treatment and the patients are followed closely for the development of MS.

Teaching pearls
- ➢ *Postcontrast fat saturation T1 images are highly sensitive for the detection of optic nerve enhancement.*
- ➢ *50–60% of patients with optic neuritis progress to MS.*
- ➢ *20% of patients with MS present with optic neuritis.*

References
Frith JA, McLeod JG, Hely M (2000). Acute optic neuritis in Australia: a 13 year prospective study. *J Neurol Neurosurg Psychiatry* **68**:246–56.
Kupersmith MJ, *et al.* (2002). Contrast-enhanced MRI in acute optic neuritis: relationship to visual performance. *Brain* **125**:812–22.
Sorensen TL, *et al.* (1999). Optic neuritis as onset manifestation of multiple sclerosis: a nationwide, long-term survey. *Neurology* **53**(3):473–8.

3 A 30-year-old male patient presents with complex partial seizures.
i. Name structures 1–3 on this coronal T2-weighted 3T MR (**3**).
ii. What are the imaging findings?
iii. What is the diagnosis?



3: Answer

3 Diagnosis
Right mesial temporal sclerosis (MTS) (**iii**).

Imaging findings
Structure 1: CA4/dentate; structure 2: subiculum; structure 3: parahippocampal gyrus (**i**). The right hippocampus is both shrunken and hyperintense on T2-weighted imaging (**ii**). This affects all parts of the hippocampal body shown, the CA1–4 areas of the cornu ammonis (see below). Other associated findings included loss of hippocampal structure and hippocampal head digitations, dilatation of the ipsilateral temporal horn of the lateral ventricle, local white matter changes, as well as atrophy of the fornix and mamillary bodies.

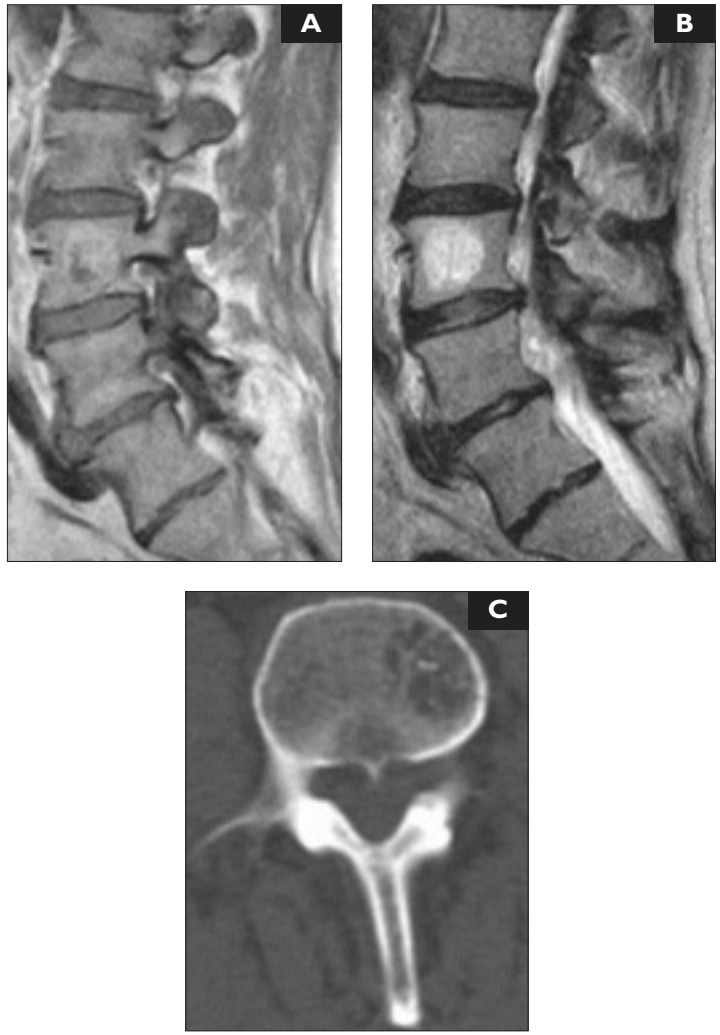
Pathology and clinical correlation
MTS is found in some patients with temporal lobe complex partial seizures and is the most common cause for epilepsy surgery. Histological examination reveals pyramidal and granule cell neuronal loss in the cornu ammonis and dentate sections of the hippocampus. The cornu ammonis can be further subdivided into 4 areas CA1–CA4. Medially, it blends into the subiculum and then onto the parahippocampal gyrus.

There is some debate over whether MTS is an acquired or developmental pathology. Temporal imaging changes showing the development of MTS have been well-documented following prolonged seizures, supporting an acquired pathophysiology. However, it has also been observed that MTS is associated with a second developmental abnormality (dual pathology) in 15% of cases.

Teaching pearls
- ➢ *The imaging findings of MTS are a shrunken, T2 hyperintense hippocampus.*
- ➢ *Underlying pathophysiology is still debated.*

Reference
Duvernoy *et al.* (). *The Human Hippocampus: Functional Anatomy, Vascularization, and Serial Sections with MRI*, 3rd edn.

4: Question



4 A patient presents with low back pain, and the following images are obtained (**4A–C**).
i. Define the level of the vertebral abnormality.
ii. What is the significance of intrinsic increased T1 signal?
iii. What is the name given to the CT appearance in **4C**?