

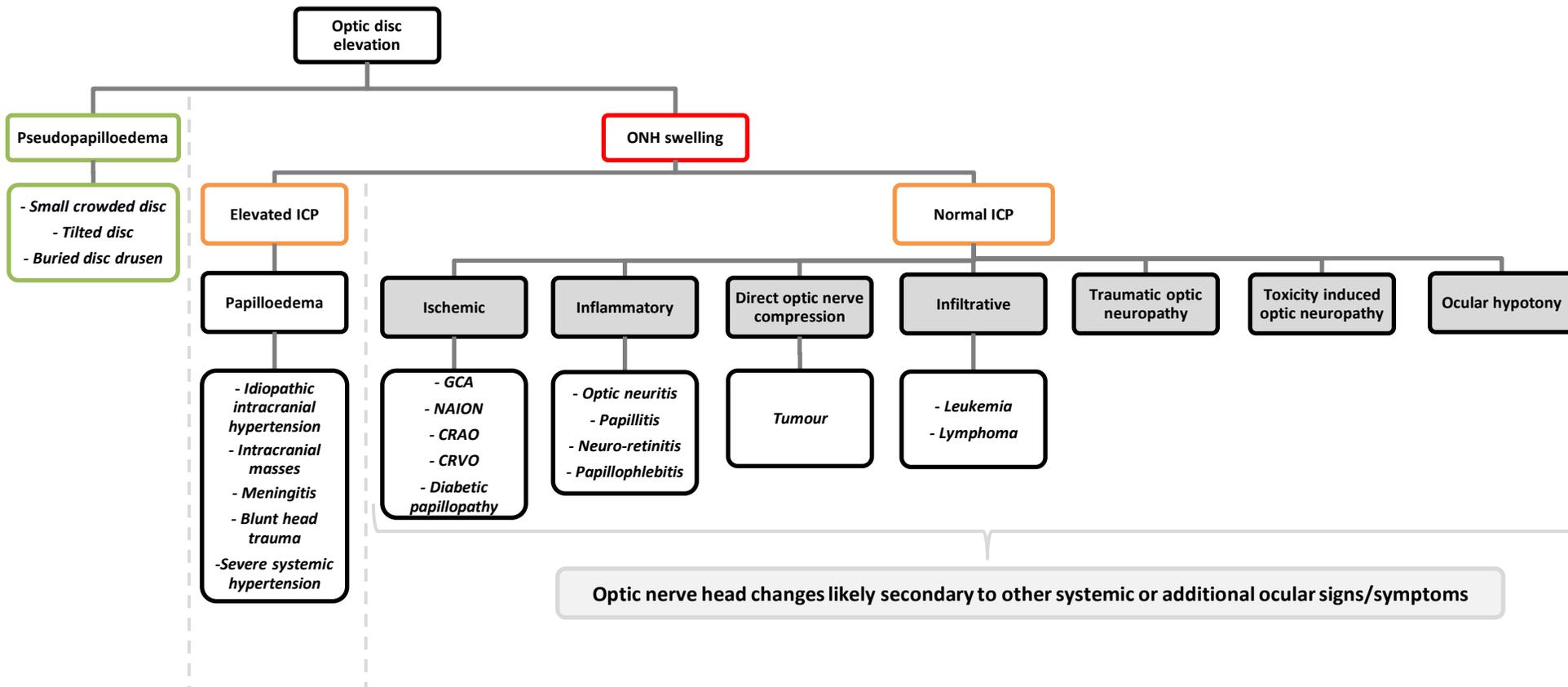
Optic disc elevation is an overlapping feature in two broad categories of conditions – pseudopapilloedema and true optic disc swelling. Accurate diagnosis of these conditions is critical as there is significant difference in urgency and implications on patient management. Pseudopapilloedema is often managed with routine review while true optic disc swelling requires urgent medical attention due to its potentially life and sight threatening consequences. The table below demonstrates the use of multimodal imaging in aiding differentiation of pseudopapilloedema and true optic disc swelling.

Differential Diagnoses of Optic Disc Elevation					©CFEH
	Colour photo	Fundus Autofluorescence (FAF)	B-scan ultrasonography	Optical coherence tomography (OCT) <i>Horizontal scan through the centre of disc</i>	Description
Pseudopapilloedema	Small crowded optic disc				
					<p><i>Small crowded optic discs are an anatomical variation caused by a normal amount of optic nerve axons or ganglion cells coursing through a small scleral foramen and converging at a small optic disc. They are commonly associated with hyperopia as a result of shorter axial length.</i></p> <p>Symptoms (Sx): Asymptomatic</p> <p>Clinical appearance: Indistinct disc margins and no apparent cup</p> <p>FAF/B-scan: Unremarkable</p> <p>OCT : Pseudo 'lumpy-bumpy' contour of the subretinal hyporeflective space due to blood vessel shadowing artefact (*). Intact adjacent retinal architecture (arrows). No visible cup</p>
	Tilted disc				
					<p><i>A tilted disc occurs when the ONH appears to enter the eye at an oblique angle. They can be congenital or acquired usually associated with myopic and astigmatic refractive error.</i></p> <p>Sx : Asymptomatic except when associated with myopic changes such as macular detachment.</p> <p>Clinical appearance: Blurred and elevated disc margins, temporal or inferonasal peripapillary atrophy (PPA)</p> <p>FAF/B-scan: Unremarkable at the disc. Hypo-autofluorescence of PPA (Δ).</p> <p>OCT : Presence of optic disc cup (*). Height difference between the surface elevation of the temporal (T) and nasal (N) aspect of the optic disc, however these can also occur superiorly and inferiorly.</p>
	ONH drusen (buried)				
					<p><i>ONH drusen are calcified deposits which may appear at the surface or buried within the disc. They may be caused by a combination of disturbance of metabolism in the axons, abnormal vasculature and a small scleral canal.</i></p> <p>Sx: Usually asymptomatic but can cause chronic peripheral vision loss. Rarely transient visual obscurations.</p> <p>Clinical appearance: Elevated disc with irregular margins. Superficial drusen are yellow & reflective.</p> <p>FAF/B-scan: Hyper-autofluorescent areas on FAF(Δ). Hyper-echoic on B-scan (arrow)</p> <p>OCT : 'Lumpy bumpy' subretinal hyporeflective space which may correspond with the contour of drusenoid deposits (*). Cysts with hyper-reflective walls (arrow).</p>
ONH swelling	Papilloedema - ONH swelling with elevated intracranial pressure (ICP)				
					<p><i>Papilloedema presents as bilateral swelling of the ONH but can be asymmetrical. ONH elevation occurs due to ganglion cell axon oedema and interstitial fluid accumulation within ONH tissue (mechanical or ischemic).</i></p> <p>Sx: Global headache worse in morning upon waking and being recumbent, transient visual loss, photophobia, dyschromatopsia, diplopia, tinnitus, nausea or vomiting</p> <p>Clinical appearance: Blurred disc margins, peripapillary RNFL thickening, tortuous vessels, dilated disc capillaries, haemorrhages, spontaneous venous pulsation loss, Paton's lines, disc hyperemia and disc pallor in chronic disease. These features can be subtle in mild disease.</p> <p>FAF/B-scan: Dense hypo-AF extending beyond the peripapillary area. Elevation of the disc on B-scan (arrow)</p> <p>OCT: Marked ONH elevation. Subretinal hyporeflective space "V contour"(*).Cystic spaces in adjacent retina (arrows)</p>
	ONH swelling with normal ICP				
	<p><i>ONH swelling with normal ICP is typically unilateral, associated with systemic changes and associated with additional ocular signs depending on the nature of the condition.</i></p> <p>Symptoms: Symptoms depend on the cause of the swelling and associated complications (see Flow Chart overleaf)</p> <p>Signs: Elevated disc with irregular margins. Other ocular signs include: retinal haemorrhages, exudates and cotton wool spots, macular oedema, uveitis, cranial nerve palsies, nystagmus and visual field defects</p>				

Flow chart of causes of optic disc elevation

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Adapted from Chiang et al, The usefulness of multimodal imaging for differentiating pseudopapilloedema and true swelling of the optic nerve head: a review and case series *Clin Exp Optom* 2015 Jan;98(1):12-24
Abbreviations: ONH, Optic Nerve Head; ICP, Intracranial Pressure; GCA, Giant Cell Arteritis; NAION, Non-Arteritic Ischemic Optic Neuropathy; CRAO, Central Retinal Artery Occlusion; CRVO, Central Retinal Vein Occlusion



Key considerations

1. Although multimodal imaging such as OCT, FAF and B-scan ultrasonography can be useful in improving the differential diagnosis of ONH elevation in clinical practice, pseudopapilloedema remains a diagnosis of exclusion. In some cases, the possibility of true ONH swelling superimposed on a small or tilted optic disc needs to be considered.
2. FAF and B-scan ultrasonography are imaging modalities which can definitively diagnose ONH drusen. Key features on other imaging modalities such as OCT should be interpreted and considered in conjunction with other clinical findings when differentially diagnosing ONH elevation.
3. Where clinical findings are inconclusive and true ONH swelling cannot be definitively ruled out, it is best to err on the side of caution and refer for further neuroimaging investigation to exclude causes of true ONH swelling.