Optic disc edema could be bilateral such as in papilledema or unilateral such as in optic neuritis or anterior ischemic optic neuropathy. However there are no strict boundaries and asymmetrical papilledema may present with unilateral disc edema and optic neuritis or ischemic neuropathy may be bilateral. If the disc edema is hyperemic, then it points towards an inflammatory cause and is classified as papillitis. However if it is pallid edema, the cause is likely to be ischemic. Both papillitis and ischemic optic neuropathy are associated with visual loss unlike papilledema where the vision is preserved till late.

The clinical presentation of patients having optic disc edema may vary from being asymptomatic or with only headache (papilledema) to severe visual loss (papillitis) depending upon the etiology of the nerve swelling.

The various causes of disc edema are described in the table below.

<table>
<thead>
<tr>
<th>Unilateral</th>
<th>Bilateral</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hyperemic disc</td>
<td>Papilledema (Figure 2)</td>
</tr>
<tr>
<td>Pale disc edema</td>
<td>Hypertensive retinopathy (Figure 3)</td>
</tr>
<tr>
<td>Minimal visual field loss</td>
<td>Anterior Ischemic optic neuropathy</td>
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<tr>
<td>Significant visual loss</td>
<td>Atypical optic neuritis</td>
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<td>Papilledema</td>
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<td>Chronic Papilledema</td>
<td>Compressive optic neuropathies</td>
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<tr>
<td>&quot;Unilateral&quot; disease involving both eyes</td>
<td>Others (uveitis, hypotony etc.)</td>
</tr>
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</table>

The most common conditions causing disc edema are discussed below:

**Anterior Ischemic Optic Neuropathy**

Anterior ischemic optic neuropathy (AION) is a general term referring to all causes resulting in temporary or permanent obstruction to the vascular
flow. It presents with segmental or sometimes diffuse pale disc edema.

There are 2 subtypes of AION, the first one being arteritic, associated with giant cell arteritis. This is a rare subtype and is associated with bilateral involvement and carries a poorer prognosis. The second one is the more common nonarteritic variety, presumably solely due to atherosclerosis and occlusive small vessel disease. It is usually unilateral though it may subsequently affect the fellow eye in a third of the cases.

**Clinical Features:**
- Unilateral, painless, moderate to severe loss of vision.
- Field defect is most commonly inferior altitudinal defect.
- Visual acuity ranges from 20/200 to no light perception.
- Age group is middle-aged to elderly.
- Non arteritic AION is usually associated with systemic vascular disease (hypertension is present in 35-50% cases, diabetes 10-25%, atherosclerosis, blood coagulopathies, nocturnal hypotension) or collagen vascular disease.
- Arteritic AION is associated with Giant cell arteritis and the patient may have associated signs of anorexia, weight loss, decreased appetite, jaw claudication, scalp tenderness and malaise.
- AION may be seen in the setting of Giant cell arteritis, Collagen vascular diseases, Syphilis, Herpes zoster, Arteriosclerosis, Diabetes mellitus, Hypertension, Embolic disease, Migraine, Sudden and severe hypotension, Cardiac insufficiency. Patients on systemic therapy with certain drugs e.g. amiodarone.

Fundus picture shows focal or diffuse pale disc edema with flame shaped hemorrhages near the disc margins. (Figure 1)

**Management**
- Complete ocular and systemic history
- Erythrocyte sedimentation rate (ESR). Elevated in arteritic but will be normal in NAION
- C - Reactive Protein (CRP)
- Temporal artery biopsy - To rule out Giant cell arteritis
- For Arteritic AION, 1-2g I.V. methylprednisolone should be given for two to three days, followed by oral steroids for two to four years to prevent vision loss from progressing to the other eye.
- For Non Arteritic AION, underlying predisposing factor (diabetes, hypertension, hyperlipidemia, smoking) should be diagnosed and treated to prevent bilateral involvement which can occur in about 33% cases.

**Optic Neuritis**
Optic neuritis (ON) is defined as acute inflammation of the optic nerve. When the inflammation involves the disc it is termed as papillitis (Figure 2). When it involves the posterior part of the optic nerve, the fundus appears normal and it is termed as retrobulbar neuritis.

**Etiologies:**
The most common etiology is demyelination, which can occur with or without evidence of multiple sclerosis (MS). Others causes include Infection (syphilis, mumps, measles), Infiltrative/ inflammatory disease (sarcoidosis, lupus) or Ischemic vascular disease (diabetes)

**Optic Neuritis and Multiple Sclerosis:**
Optic neuritis (ON) is the initial presenting sign in 20 to 25 percent of MS patients and occurs in about 50%
of cases of MS. About 35% to 75% of patients who present with ON go on to develop clinical MS. Although long term studies from the Asian population are not available, the risk of progression to clinical MS appears to be low. The risk of developing MS increases steadily during the first 10 years after the initial presentation of ON.

**Clinical features:**
The clinical presentation of demyelinating optic neuropathy varies.
- Sudden onset, moderate-severe loss of vision, which can be progressive for about 10-14 days and then will stabilize and should start improving.
- Periocular eyeache, increasing on eye movements.
- Dysschromatopsia.
- Decreased brightness sense.
- Relative afferent pupillary defect in the involved eye.
- Visual field defects (could be of variable severity and type though frequently central / cecocentral).
- Disc swelling with or without vitreous cells.
- Uhtoff’s sign (decreased vision with or without limb weakness following increasing body temperatures i.e., a bath or exercise),
- Romberg’s sign (patient falls when they close their eyes),
- Pulfrich’s stereo phenomenon (beer barrel appearance to the environment).
- Systemic signs and symptoms may include headache, nausea.

**Management**
The currently accepted management is with intravenous methylprednisolone sodium succinate 250 mg every 6 hours or 1 gram every day for three days followed by oral prednisone (1 mg/kg per day) for 11 days as per the Optic Neuritis Treatment Trial (ONTT). At our centre we are routinely giving intravenous dexamethasone 200 mg every day for three days followed by oral taper. The aim of this treatment is for the purposes of accelerating visual recovery only and it does not affect visual outcomes after one year. The ONTT also determined that the use of oral prednisone in routinely prescribed doses (1 mg/kg per day) alone for 14 days is contraindicated and was associated with increased risk of reoccurrence. Patients receiving this therapy had a higher rate of new attacks of ON in both the initially affected and fellow eyes than did the group receiving intravenous steroids and placebo.

Treatment with intravenous methylprednisolone followed by oral corticosteroid regimens also reduces the two-year risk of development of clinical MS, particularly in patients with demyelinating lesions on MRI of the brain at the time of episode of ON. Though serious side effects of dexamethasone or methyl-prednisolone therapy are infrequent, monitoring during I/V administration is necessary. Prior to giving steroids, a chest X-ray and proper clinical examination to rule out tuberculosis or any other focus of infection is necessary.

**Papilledema**
Papilledema is an optic disc swelling that is secondary to elevated intracranial pressure (Figure 3). In contrast to other causes of optic disc swelling, vision usually is well preserved with acute papilledema. Papilledema almost always presents as a bilateral phenomenon though may be asymmetrical and may develop over hours to weeks. The term should not be used to describe optic disc swelling with underlying infectious, infiltrative, or inflammatory etiologies.
Papilledema will occur only if there is continuation of the subarachnoid space of the brain with the spaces around the optic nerve sheath. Papilledema does not occur in eyes with pre-existing optic atrophy.

**Clinical features:**
- H/o nausea, vomiting, headache, nerve palsies etc.
- Usually Bilateral
- No significant visual function loss in the early stages
- Transient obscuration of vision
- Enlarged blind spots and peripheral field constriction
- Diplopia due to VI nerve underaction

**Stages of Papilledema:**
**Early**
- Hyperaemia, indistinct margins (lower pole first)
- Blurring of peri-papillary retinal nerve fiber layer
- Splinter haemorrhages in the peri-papillary region
- Absence of spontaneous venous pulsations (absent in 20% of normals)

Established
- Obvious swelling, numerous haemorrhages
- Engorged veins, obscuration of surface vessels
- Paton’s lines, cotton wool spots, macular fan or star

Chronic
- Haemorrhages and exudates resolve
- Disc has rounded appearance, milky gray colour
- Obliteration of cup, RNFL defects (slit like)

Atrophic
- Pale disc
- Narrow and sheathed vessels
- Peri-papillary pigmentary changes / choroidal folds

Etiology:
Any tumors or space-occupying lesions of the CNS
Idiopathic intracranial hypertension (IIH)
Decreased CSF resorption (eg, venous sinus thrombosis, inflammatory processes, meningitis, subarachnoid hemorrhage)
Increased CSF production (tumors)
Obstruction of the ventricular system
Cerebral edema/ encephalitis
Craniosynostosis

Management:
The diagnosis of the cause would include the need for certain tests including Hemogram, Chest X-ray, MRI or CT scan of the head and a lumbar puncture.
Treatment essentially involves treating the underlying cause such as an intracranial space occupying lesion.

Management of Idiopathic intracranial hypertension (IIH):
The treatment of IIH is initially medical including weight loss, use of Diuretics (Lasix, furosemide) and acetazolamide. If these fail then surgical therapy in the form of Optic nerve sheath fenestration or ventriculo peritoneal shunts may be considered.

Pseudopapilledema
- Usually associated with hyperopia.
- Small disc, with absent physiological cup.
- Abnormal branching and tortuosity of retinal vessels. No dilated capillaries on surface of the disc.
- Increased number of central retinal vessels arising from optic disc.
- Scalloped margins/ irregular disc surface or visible optic disc drusen.
- Not associated with any retinal haemorrhages, exudates, cotton wool spots.
- On FFA: may see autofluorescence at disc. No disc leak.
- USG may show high spikes at disc even with low gain settings due to presence of drusen.

Compressive optic neuropathies
- Lesion causing local compression of the optic nerve in the orbit or just retrobulbar may present with disc edema.
- Transient monocular visual loss, in a particular gaze (direct pressure of optic nerve or interruption of blood supply).
- Enlargement of blind spot on field analysis.
- RAPD and color vision abnormalities.
- Chronic compression leads to a triad of visual loss, swelling evolving into atrophy and optociliary shunt (shunting the blood from retina to choroid).
- Rarely optic disc swelling may occur in intracranial lesions eg. Sphenoid wing meningeoma.
- Features of orbital disease: proptosis, limitation of ocular movements, orbital congestion.
- Diagnosis can be made with the aid of ultrasound of the orbit or a CT or MRI.
- Essential to rule out thyroid ophthalmopathy and pseudotumor.

Although disc edema may be due to a large number of causes, a proper history, detailed systemic and ophthalmic examination and a systemic approach can lead to the correct diagnosis in a majority of cases and avoid unnecessary and expensive investigations in most.