THE NORTHERN HOSPITAL

CAROTID AND VERTEBRAL ARTERY DISSECTION

"Nell Gwynn", oil on canvas, c.1680 , Sir Peter Lely.

"…Miss Nelly has been dying of an apoplexy. She is now come to her sense on one side, for the other is dead of a palsy…"

Sir Charles Littleton, letter to his friend, November 1687.

The woman depicted in this 17th century portrait caresses her pet lamb, a common symbol at the time of "innocence". Yet if so innocent, why the exposed breast? There is a hidden message here. This is the notorious Nell Gwynn, the most famous stage actress of her time and most beloved of all King Charles II's numerous mistresses. She was possibly a prostitute in her early career, yet remained fiercely loyal to Charles to the end of his life, once he took her under his "patronage". She died at just 37 years of age from a stroke. Even in the 17th century death from a stroke was unusual at such a young age. Some believed at the time she had contracted syphilis from the King. Young people like Nell, who present with a stroke, are most unlikely to have suffered it as a result of atherosclerotic disease. Like Sir Peter Lely's depiction there is likely to be hidden "pathology" in these cases. Whilst syphilitic vasculitis was far more common in the 17th century, carotid artery dissection should be considered in the 21st century.

CAROTID AND VERTEBRAL ARTERY DISSECTION

Introduction

Cervical artery dissections are dissections of the **carotid** or **vertebral** arteries.

Carotid and vertebral artery dissections are significant causes of **stroke** in **young people**.

While any of the arteries in the neck may be affected, **internal carotid artery dissections** are by far the most common. **Vertebral artery** dissections are less common.

Both types can occur intracranially and extracranially.

Intracranial involvement is rarer but is more serious.

Causes can be **traumatic** or **spontaneous.**

The investigation of choice is 4 vessel cervical CT angiography.

Treatment options include:

- Antiplatelet therapy
- Anticoagulation with heparin
- tPA (in carefully selected cases).
- Endovascular interventions.

Epidemiology

Cervical artery dissections can occur in all age groups

They are a significant cause of stroke in **young people** (< 45 years).

Carotid artery dissections are most common in young adults.

While the mean age for extracranial internal carotid artery dissection is 40 years, intracranial dissections are more common in those aged 20-30 years.

Approximately 20% of strokes in the young are caused by carotid artery or vertebral artery dissections in the neck, compared to only 2.5% in older patients.

There is an increased incidence with a positive **family history** for this disease.

Pathophysiology

Mechanism:

Dissections of the carotid and vertebral arteries usually arise from an initial intimal tear.

The tear allows blood under arterial pressure to enter the wall of the artery and form an intramural hematoma, or "false lumen".

The intramural hematoma is located within the layers of the tunica media, but it may be eccentric, either toward the intima or toward the adventitia.

A subintimal dissection tends to result in stenosis of the arterial lumen, whereas a subadventitial dissection may cause aneurysmal dilatation of the artery.

Embolism from thrombus formation at the dissection site is thought to play the major part in stroke pathogenesis. This is supported by Transcranial Doppler studies showing cerebral microemboli soon after dissection and by brain imaging results suggesting an embolic pattern. [3]

Causes:

Causes include:

1. Traumatic:

    - **This can be minor or major, blunt or penetrating.**

      Cause can include:

      ♥ Direct injury

      ♥ Seemingly minor injury:

Dissection may even follow *relatively minor* trauma, such as attempts at neck manipulation. Presumably dissection occurs in these cases in the *predisposed*.

- ♥ "Whiplash" type injury
- ♥ Strangulation

2. Spontaneous:

- Suspect an underlying vascular pathology such as:
  - ♥ Marfan's syndrome
  - ♥ Fibrocystic dysplasia
  - ♥ Arteritis/ connective tissue diseases.
- Hypertension.
- Idiopathic:
  - In a number of cases the causative factor remains unclear.

Complications of dissection:

These include:

1. Vessel occlusion, leading to stroke.

2. Thrombus formation with embolism, leading to stroke.

- The dissection may cause the formation of a thrombus from which fragments may embolize.
- TIAs are thought to be the result of transient hemodynamic instability, whilst completed stroke is thought to be due primarily to embolization.

3. Vessel rupture:

- Intracranially this results in subarachnoid hemorrhage.

4. Pseudoaneurysm formation

Morbidity from carotid artery dissection varies in severity from transient neurologic deficit to permanent deficit and death.

Intracranial internal carotid artery dissection is associated with a high mortality rate (75%).

The risk of stroke is greatest in the first 24 hours after cervical artery dissection and decreases over the next 7 days. The risk of stroke after 2 weeks is relatively low.

Prognosis

In general terms:

- Prognosis of carotid artery dissection depends on the severity of the initial ischemic episode, the degree of collateral circulation and the site of the dissection.
- Prognosis is more favorable for extracranial dissection than for intracranial dissection.
- *Overall*, the prognosis is fairly good in cases of *spontaneous* extracranial dissection, with recanalization often occurring within 7-30 days.

Clinical features

Carotid and vertebral artery dissection can be very difficult diagnoses to make.

Patients may present as stroke or TIA in a young person where the diagnosis is easier.

Dissection however may also present initially as vague headaches, facial pain or neck pain where diagnosis is far more problematic.

The following clinical features may occur:

Important points of history:

1. Pain:

   There is a history of unexplained, **facial** or **neck (particularly over the course of the carotid artery) pain** or **headache.**

   Note that pain may precede a cerebral ischemic event, TIA (including amaurosis fugax) or completed stroke, in the carotid artery distribution, by a period of hours or even days.

   The pain will usually be experienced ipsilateral to the side of the dissection.

   The precise location of pain is neither sensitive nor specific for the artery of dissection, however in *general terms:*

   - Headache:
     - ♥ Pain from a carotid dissection tends to present as a frontal headache.
     - ♥ Pain from a vertebral artery dissection tends to present as occipital headache.

- Neck pain:
  - ♥ Carotid dissections result in anterior neck pain
  - ♥ Vertebral dissections result in posterior neck pain.
- Facial pain:
  - ♥ Eye, ear, or face pain is very likely to indicate carotid involvement.

2. Neurological symptoms:
   - Patients may present complaining of overt neurological signs, such as hemiparesis or monocular visual disturbances.
3. History of trauma:
   - A history of trauma of course makes traumatic dissection likely.

     However when the trauma is mild, trivial or even unrecognized (as in the case of cervical manipulations), then diagnosis is far more problematic.

Important points of examination:

1. Assess for any neurological deficits:
   - Hemiplegia/ hemisensory loss
   - Unilateral cranial nerve deficits may be associated with brainstem ischemia
   - Monocular visual disturbances
2. Vascular bruits:
   - Only about one-third of patients with carotid dissections have bruits.

     Therefore the absence of a carotid bruit does **not** exclude the possibility of an arterial dissection.
3. Ipsilateral neck tenderness may occur:
   - Recurrence of neck pain and tenderness may indicate an extension to the dissection.
4. Ipsilateral Horner's syndrome:
   - A strong but subtle clue to the condition is the presence of **Horner's syndrome** in association with ipsilateral neck pain and tenderness.

This is presumably due to a sudden expansion of the internal carotid artery with compression of sympathetic nerve fibers.

See also separate document on Horner's syndrome in Neurology folder

5. Setting of trauma:

Clinical findings that raise suspicion for a vascular injury in the setting of trauma include:

- Expanding neck hematoma
- Hemiplegia in the alert patient, and the development of a depressed level of consciousness in the patient who was initially lucid, without obvious head injury, or who had a normal head CT.

  The significance of the lucid interval with deteriorating level of consciousness has been appreciated by many clinicians and points to the development of cerebral ischemia secondary to vascular injury.

  These patients require emergent evaluation of **all 4 cervical vessels**, either through CTA, 4 vessel angiography, or direct surgical exploration.

Investigations

Blood tests:

1. FBE
2. ESR (if vasculitis is suspected or in cases of apparent spontaneous dissection).
3. CRP (if vasculitis is suspected or in cases of apparent spontaneous dissection).
4. U&Es/ glucose.
5. Clotting profile.

ECG:

AF, looking for a possible alternative cardiac source of emboli.

Carotid Doppler Ultrasound:

Carotid Doppler ultrasound is useful as an initial **screen**, especially where MRA or CT angiogram is not available.

Significant limitations of ultrasound scanning however include:

- Inability to image the distal internal carotid artery and intracranial arteries.

- Inability to image the vertebral vessels
- Lower sensitivity compared to CT angiogram or MRA

CT Scan Brain/ CT angiogram:

CT angiogram (full 4 vessel - carotid arch to vertex) is the best initial investigation.

This will also rule out possible associated intracranial hemorrhage.

MRI / MRA:

MRA/MRI is an alternative to CTA, when this is contraindicated to a significant contrast allergy

Digital Subtraction Angiography:

Digital subtraction angiography remains the "gold standard" of imaging modalities.

In addition to clear images, angiography also provides a better assessment of blood flow than CTA or MRA imaging. Angiography can provide information on collateral blood flow, and this may guide further management and stratify risk. Patients with an intact circle of Willis should tolerate an acute occlusion much better than those who do not.

With the ready availability of **CTA**, DSA is rarely required, unless the CTA is equivocal, and strong clinical suspicion remains.

Management

Treatment is difficult and not standardized.

It will depend on a number of factors, including:

1. The cause:
    - Trauma versus spontaneous
2. The anatomic location of the lesion:
    - Carotid versus vertebral
    - Intracranial versus extracranial
3. Complicating factors:
    - Associated hemorrhage

Options include:

1. Antiplatelet therapy/ anticoagulation:

- Antiplatelets and anticoagulants in the first three months after the stroke have a **similar** benefit and risk profile when used in the management of cervical artery dissection., (although the current quality of evidence for this is only moderate) [3]

  Antiplatelets may be preferred given the perception of lower risk and potentially easier adherence due to single daily dose and no need for blood test monitoring. [3]

2. Endovascular interventions:

   - Endovascular therapies (such as stenting) for extracranial carotid or vertebral artery dissection may be an option when medical management fails or is contraindicated.

     Endovascular therapy may also reduce the risk of stroke by stenting the dissecting vessel. Aneurysms complicating dissections can be filled with soft coils to induce a thrombosis. This method is a definitive treatment of aneurysms, and long-term rates of rupture are very low.

     Initiation of medical therapy with antiplatelet therapy, or anticoagulation does not exclude patients for endovascular therapy; however, when the medical therapy fails it may be too late to prevent neurological deficits.

3. Trauma:

   - In the setting of trauma aggressive screening protocols for dissection will identify increased numbers of cervical dissections before development of stroke symptoms.

     Screening should not wait for the development of neurologic signs or symptoms in those at significant risk.

     It is hoped that early detection will reduce the incidence of stroke and death in the setting of traumatic dissection.

Disposition

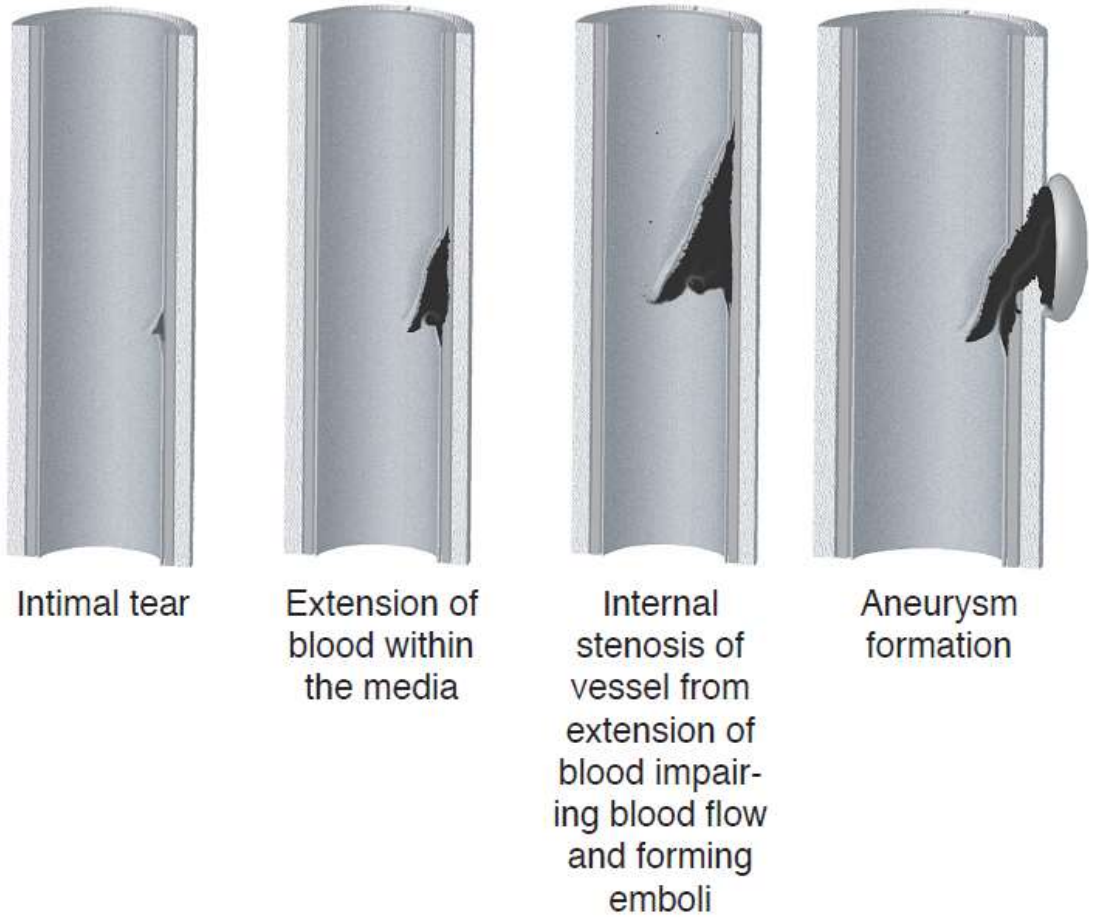
Trauma patients will usually be admitted under a **Trauma Unit** with **Vascular Unit input.**

Patients with spontaneous dissections will usually be admitted under a **Stroke Unit.**

Patients who may be suitable for endovascular intervention are referred to the **Vascular Unit.**

Appendix 1

Mechanism of arterial dissection:



The arterial wall essentially consists of three layers:

- *Intima*
- *Media*
- *Adventitia*

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References:

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2. Kenneth Shea et al; Carotid And Vertebral Arterial Dissections in the Emergency Department. Emergency Medicine Practice, Volume 14, Number 4, April 2012.

3. Clinical Guidelines for Acute Stroke Management - National Stroke Foundation Website, (accessed November 2017):

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