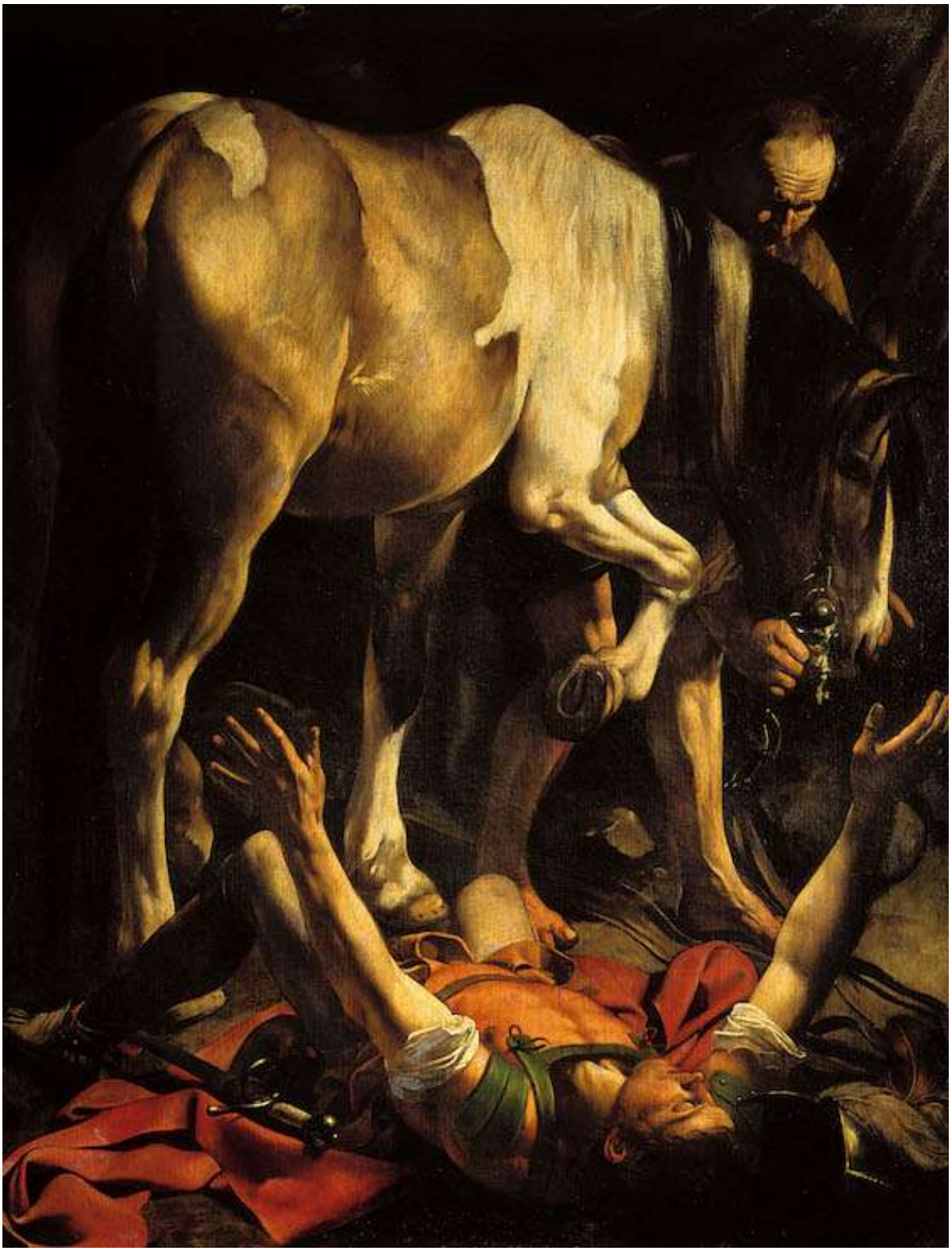
THE NORTHERN HOSPITAL

AMAUROSIS FUGAX



"The Conversion of St. Paul", oil on canvas, 1601, Michelangelo Merisi da Caravaggio.

All the instincts of post-Renaissance art were to respond to spatial confinement by using perspective to open up the space. With the theme of Mary's ascent into heaven, Annibale had naturally followed this precept, so his painting was full of bright light. But Caravaggio's best moments were often counter-intuitive. Instead of trying to defeat the cramped room optically with smaller figures and illusions of deep space he did exactly the opposite, bringing his massive figures of men and beasts right to the very picture edge so that, immense and lumbering, they seem, alarmingly, about to fall into our personal space….Instead of relief and depth, we get holy claustrophobia…How it works! Witness was never closer, the distance between observer and event never more successfully annihilated…

…Caravaggio has a moment of blinding insight of his own, parting with centuries of Pauline iconography, including his own earlier version of the saint, as the usual bearded elder. Caravaggio thinks instead of the local cops; young, brutal, stubble-chinned, strapping swaggerers like the bravi he fell foul of all too often. Taking years off Paul, of course, only makes the stunning force of the light that had thrown him all the more potent. It's a literally dazzling concept.

Once again Caravaggio isn't separating his own life from art, but rather bringing his gift for raw-boned heft to his one-man revolution in sacred painting. He uses the narrow space and our viewing angle to force us down, as if he were pushing us bodily to the ground (he had lots of practice) so that we find ourselves beneath the piebald's raised hoof. Instead of the routine angels that crowded his first effort, there are just three characters: horse, groom and floored apostle. But as with "The Calling of Saint Mathew", the shock of the painting is in the light that floods over the horse's body and bounces back from Paul's torso and face and so that its strength reflects in the heavily veined leg and creased forehead of the gentle groom, enfolding him too in its saving illumination. The attributes of Paul's worldly power are broken: plumed helmet thrown off, armour straps unfastened, and the eyes that had sought out Christians to harass are now an extraordinary yellow, as if the cornea had first been scorched by the light, then covered, as the Gospel says, with a filmy cataract. Blinded, he will in three days, be rewarded with the first true sight of his life.

Simon Schama, "The Power of Art", BBC, 2006

The causes of transient visual loss are legion. In a religious age, Caravaggio painted the most important cause - a blinding epiphany.

In the modern age of medical science, we must take a Caravaggian view of amaurosis fugax - in the technical sense the causes are legion; but our epiphany must be for the main game - and these are the vascular causes.

AMAUROSIS FUGAX

Introduction

Amaurosis Fugax is generally considered to be a **particular type** of **anterior circulation** transient ischaemic attack which involves acute transient monocular loss of vision due to retinal ischemia.

There is some confusion in terminology, but **binocular hemianopic** loss of vision due to a posterior circulation disorder is more often considered as a "traditional" form of TIA, rather than being labelled as amaurosis fugax.

Some literature refers to amaurosis fugax as transient visual loss due to *any* cause. This is probably unhelpful as a vast number of conditions will distract the clinician from the most important vascular issues of high risk TIA and high risk for visual loss. Additionally, the generalized term becomes almost meaningless with regard to standardizing investigation and treatment protocols. The following refers to amaurosis fugax from **vascular pathology.**

Anterior circulation TIAs are routinely risk stratified in the ED according to the **ABCD2** system.

All cases of monocular (vascular) amaurosis fugax, however are *immediately* classified as high risk as there is a strong association with high grade carotid artery stenosis.

Its high importance lies in:

- The high risk of subsequent stroke in general.
- The risk of recurrent episodes resulting in permanent loss of vision in particular.

All cases of amaurosis fugax, should be admitted to a stroke unit (or a short stay unit with specialist neurologist consultation, according to local practice) in order to fully investigate the patient in an appropriately timely manner.

See also separate documents on:

- **TIA (in Neurology folder).**
- **Stroke: Infraction (in Neurology folder).**
- **Central Retinal Artery Occlusion (in Ophthalmology folder).**

History

The terminology "amaurosis fugax" was derived from the Greek "amaurosis", meaning dark, and the Latin "fugax" meaning fleeting - and so "fleeting darkness".

Pathology

Causes:

Causes include:

1. Arterial atherosclerotic disease:
    - Emboli from carotid artery disease, (most commonly)
    - Emboli from a cardiac source (less commonly)
    - Emboli from more distal vessels, including the Internal Carotid or Ophthalmic artery, (uncommon).
    - Transient hypoperfsion from a stenotic artery, may also be a cause.

Uncommonly:

2. Procoagulation disorders
3. Vasculitic disease:
    - ♥ In particular Giant cell (or "temporal" arteritis), in patients > 50 years.
4. Migrainous vasospasm.

Monocular versus Binocular Visual Loss:

Transient **monocular** visual loss implies a disorder **anterior** to the optic chiasm (i.e, the optic nerve or the eye/ retina). This may be due to intrinsic ocular disease or vascular ischemia due to the ipsilateral **anterior circulation** - i.e carotid or carotid branch arterial disease.

Transient **binocular** visual loss suggests a **posterior circulation** disorder, involving the optic chiasm, tracts, or radiations, or the visual cortex. This may be due to vertebrobasilar TIA or migraine.

Clinical assessment

In the ED the patient is assessed for:

- Their vision
- Their CVS disease risk profile
- A possible embolic source for their symptoms.

Important points of history:

1. Assess the nature of the visual loss:

   In amaurosis fugax visual loss is:

   - Monocular:

     ♥ Implying a disorder *anterior* to the optic chiasm, (i.e. a lesion of the eye or of the optic nerve).

     Transient *binocular* visual loss suggests a more *posterior* process, involving the optic chiasm, tracts, radiations, or the visual cortex itself.

     In practice, however many patients are not able to state definitively whether the episode affected one or both eyes. When the history is unclear, it is best to assume that *either* a monocular or binocular aetiology is possible.

     ♥ The visual loss can be a mild blurring or fogging to complete blackness, and may it may involve a part of or all of the visual field.

   - Painless:

     ♥ Thromboembolic amaurosis fugax is painless.

     If pain is significant then migraine, angle closure glaucoma, and giant cell arteritis, need to be considered.

   - Transient:

     ♥ Typically impairment is only brief, lasting seconds to minutes, but occasionally may last one to several hours.

2. CVS disease risk assessment:

   - Assess the patient's general risk profile for CVS disease, most importantly: hypertension, diabetes, high blood lipids, previous history of CVS disease or PVD.

     When the clinical presentation is ambiguous or unclear, then the **individual risk profile** that a patient carries will largely determine the *index of suspicion* for attributing the patient's symptoms to a true episode of amaurosis fugax.

Important points of examination:

1. Document the visual acuity in both eyes
2. Check for visual field defects.
3. Assess the fundus of the eye:
    - Apart from any pre-existing disease, this will usually be normal, although an embolus in a retinal arteriole may sometimes be seen.
4. Search for a possible embolic source:
    - Check for AF
    - Carotid bruits:
        - ♥ This can be documented, but is an unreliable sign, as its presence or absence does not rule in or rule out a significant stenosis.
    - Cardiac murmurs:
        - ♥ Possible valvular lesion.

Differential Diagnoses:

The following may also explain *transient monocular* loss of vision:

1. Optic disc demyelinating diseases, (MS).
2. Larger vitreous "floaters" may occasionally obscure central vision.
3. Migraine variants, (migraine usually causes a hemianopia).
4. Transient central retinal venous occlusions.

Investigations

Blood tests:

1. FBE
2. U&Es/ glucose
3. ESR / CRP:
    - For **temporal arteritis** or other vasculitic cause.

5. Clotting profile

6. Procoagulation screen

7. Fasting blood lipids profile.

ECG:

In particular for AF, as a possible embolic source.

A period of halter monitoring may also be considered, if transient arrhythmias are suspected.

Carotid Doppler Ultrasound:

This is a useful screening test for the detection of significant carotid artery stenosis.

CT Scan/ CT Angiogram of the head and neck arteries:

Plain CT scan has limited utility in the setting of amaurosis fugax, other than to rule out an acute intracerebral bleed, particularly when the clinical presentation is somewhat unclear.

Ruling out an intracerebral bleed will also allow for the commencement of antiplatelet or anticoagulant therapy if required.

CT Angiogram is a much better investigation. It directly visualizes arterial vascular lesions of the carotid and vertebrobasilar systems, and can be immediately utilized when MRA/MRI is not readily available.

MRI/ MRA:

This represents the best imaging modality for visualizing and assessing the carotid and vertebrobasilar vascular systems.

It is also a good option in those patients not suited to the IV contrast of a CT angiogram, (allergic reactions or significant renal impairment).

Echocardiography:

The need to do this will be based on the index of suspicion for a cardiac source of embolism.

It may also be considered in younger patients in whom no obvious cause has been found,

Management

1. Antiplatelet therapy:

Options include:

- Aspirin
- Clopidogrel
- Asasantin, (dipyridamole and aspirin).

2. Anticoagulation, (heparin, clexane, warfarin):

   - This should be considered when there are recurrent/ crescendo symptoms despite antiplatelet therapy or when there is a clear cardiac source, such as *atrial fibrillation.*
   - Bridging heparin or clexane should be considered in these high risk scenarios, following consultation with the Neurology or Vascular units (as clinically indicated), followed by warfarin therapy.

3. Control of CVS risk factors:

   - This is important where these risk factors exist

4. Surgery:

   - The higher the grade of stenosis, the greater will be the benefit from carotid endarterectomy.

     Carotid artery endarterectomy is usually recommended, in patients suitable for surgery, who have amaurosis fugax and a **moderate (50-70%)** or a **high grade (> 70%)** carotid artery stenosis.

Disposition

All cases of amaurosis fugax, should be admitted to a stroke unit (or a short stay unit with specialist neurologist consultation, according to local practice) in order to fully investigate the patient in an appropriately timely manner.

Those with moderate or high grade carotid artery stenosis should be referred to the **Vascular Unit.**

Ophthalmology consultation may also be warranted where the cause of the symptoms is unclear.

References

1. Clinical Guidelines for Acute Stroke Management National Stroke Foundation 2010.

2. Syndee Givre et al, Amaurosis Fugax in Up to Date Website, 31 January 2014.

Dr J. Hayes

Acknowledgements:
Dr Jorge Zavala.
Mr Franklin Pond.
Reviewed April 2016.