**Amaurosis Fugax: A Clinical Comparison**

BY DAVID H. SLEPYAN, M.D.,* ROBERT M. RANKIN, M.D.,† CHRISTOPHER STAHLER, JR., M.D.,‡ AND GERALD E. GIBBONS, M.D.‡

Abstract: Amaurosis Fugax: A Clinical Comparison

- Amaurosis fugax (transient monocular blindness) is a symptom of retinal ischemia just as contralateral hemiparesis and sensory loss are symptoms of cerebral ischemia. These symptoms are produced by atherosclerotic stenosis of the carotid vessels at the ipsilateral carotid bifurcation and emboli from these areas causing focal, repetitive, retinal ischemia.

A study of 31 endarterectomy patients was undertaken to see if eight patients with amaurosis fugax (25%) could be differentiated from 22 patients with transient cerebral ischemia. The patients with amaurosis fugax were found to be younger. They all had 75% or greater stenosis of the internal carotid artery at the bifurcation on the symptomatic side. They all had unilateral visual symptoms and these symptoms were relieved by surgery. The patients with amaurosis fugax were devoid of cardiac disease, while 45% of the cerebral ischemic patients had documented myocardial disease.

Amaurosis fugax (transient monocular blindness) in the setting of clinically significant atherosclerosis of the carotid vessels is an indication for carotid endarterectomy.

Additional Key Words: carotid endarterectomy, retinal ischemia, emboli, cerebral ischemia, internal carotid stenosis, atherosclerotic cardiac disease

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Pathophysiology

Reviewing the natural history of amaurosis fugax, Marshall and Meadows⁵ found that neither frequency, duration, mode of onset, nor number of attacks gave any clue as to which patients would incur severe complications. They followed 80 patients with amaurosis fugax of varying etiologies over a six-year course and 16% progressed to monocular blindness or hemiplegia. These authors concur that carotid angiography is indicated in any patient presenting with these symptoms.

Nonatheromatous disease must be considered in the pathophysiology of amaurosis fugax. In most series, at least 10% of patients will be free of clinical atherosclerosis. The differential diagnosis in these patients may include giant cell arteritis, lupus erythematosus, malignant hypertension, migraine, Reynaud’s disease, sickle-cell anemia, hypoglycemia or polycythemia vera. There is even a small group of young individuals in whom no cause may be identified.⁶

In 1905, Chiari⁴ described thrombi in intracranial branches of the internal carotid which arose from mural thrombosis of an atheromatous plaque at the carotid bifurcation. He emphasized that disease of the carotid bifurcation was the main source of emboli causing strokes in elderly patients.

These observations were confirmed by Fisher⁶ when he visualized emboli in the retinal vessels of patients undergoing attacks of amaurosis fugax. Similar emboli have been visualized in fundi of patients after carotid angiography and carotid endarterectomy.⁶ This presumably results from mechanical dislodgement of atheromatous material, thrombi or platelet fibrin aggregates.

The emboli occlude the retinal vessels transiently, and those consisting of platelet fibrin aggregates are rapidly lysed and passed onward. Atheromatous debris consisting of calcific cholesterol fragments may occlude vessels causing permanent visual field

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‡This paper was supported in part by the Wenatchee Valley Clinic Medical Research Foundation, Wenatchee, Washington.
The use of an internal shunt serves two purposes and conver-
tumes of symptoms so commonly described by these patients. The repetitive nature of these attacks is  best explained by streaming of emboli from carotid  atheromata with apparent uniformity of embolic size. The tight stenosis of the internal carotid artery seen in these patients predisposes to mural thrombus formation within the internal carotid artery. Fragments of the thrombus may cause embolic occlusion of the ophthalmic artery showing multiple platelet fibrin aggregates into retinal vessels in a repetitive fashion. Vasospasm and critical blood flow to the eye are tenuous explanations for these attacks. In the case of vasospasm, ocular vessels are among the most unreactive in the body to pressure and humoral influences. Critical blood flow to the eye does not explain the symptoms of amaurosis fugax, for these attacks are known to occur without antecedent hypotension. Systemic hypotension could explain localized ischemia beyond a segmental stenosis in cerebral vessels if collateral circulation was borderline. This does not explain repetitive attacks, however.

We are left with a mechanism based on the fundamental atherosclerotic arterial lesion and an embolic event capable of changing focal cerebral blood flow rapidly. The localized stenosis of the internal carotid artery at its origin and emboli (either platelet fibrin aggregates or cholesterol debris from ulcerating plaques) give rise to repetitive transient cerebral or retinal ischemia.

Methods

Our study consists of 31 patients who were evaluated over a five-year course. Eight of these 31 patients presented with the sole complaint of transient unilateral loss of vision. Patients who were candidates for surgery were individually reviewed and referred by the neurologist in our group. His analysis of symptoms and radiological findings were used for the sake of consistency in the study. All patients underwent neurological evaluation, angiographical studies, EEG or brain scan, and postoperative Doppler studies of the extracranial vessels. Each patient in the study underwent carotid arteriography or four-vessel arch studies, as deemed necessary by clinical or radiological findings.

Endarterectomy of the carotid bifurcation was carried out with the use of an internal shunt, in all cases except one. The use of an internal shunt serves two purposes and contributes to the relatively low morbidity and mortality of the series. (1) The shunt maintains and gives increased perfusion to the symptomatic cerebral hemisphere during surgery. (2) The use of occlusive tapes and the shunt localize the area of dissection allowing more time for meticulous debridement and removal of debris. This prevents, by careful back bleeding, embolic debris from transversing the internal carotid artery. Showers of debris to the retina during endarterectomy have been demonstrated and are painfully reproducible. Any method by which iatrogenic emboli are reduced results in an improved postoperative course.

We do not feel there is a need for special hypobaric or hypercarbic adjuvants to anesthesia. We strive to maintain the patient in a normotensive state during anesthesia with adequate oxygenation to the tissues and have found standard methods completely adequate for maintaining these patients intraoperatively.

Criteria for Selection of Patients

Our indications for cerebral angiography include: all patients with focal TIAs (focal neurological deficit of cerebral vascular origin lasting for 24 hours or less, including AF), patients who have periodic transient confusion but not with transient vertigo, those patients having TIAs lasting longer than 24 hours but with complete recovery, and those patients who recovered from completed infarction to a functional self-care capability.

Contraindications to angiography consisted of advanced physiological age (80 years maximum), myocardial infarction in the preceding six months, and severe non-neurological disease (e.g., uncontrolled diabetes, severe liver disease, severe chronic obstructive pulmonary disease or chronic cardiac failure). Hypertension and controlled diabetes were not a contraindication to angiography. Most of the procedures were retrograde femoral catheterization giving full four-vessel arch studies. This technique allows selective views and deals with the common bilaterality of the disease process.

One hundred and forty-five patients were selected for angiography by the above criteria, and of these 41.4% had significant lesions (see Results). Approximately half of these patients with significant lesions underwent endarterectomy. The unoperated half consisted of those who refused surgery, had multiple extracranial stenosis or occlusion, or limiting intracranial stenosis. Patients were selected for endarterectomy who had transient ischemia or amaurosis fugax without permanent deficit and documented significant lesion by angiography. Patients with occlusion of the internal carotid vessels, or with completed strokes with severe deficit, or patients with prolonged periods of confusion in addition to symptoms of cerebral ischemia were not recommended for surgery. These exclusions were because of uniformly poor results in other series with this type of patient.

Results

Mean age of the eight patients with amaurosis fugax was 60 years, while the mean age for the 22 cerebral ischemia patients was 65 years (table 1). There were 21 men and 10 women in the total series, 2.5 to 1 ratio of men to women, which is about the same distribution

<table>
<thead>
<tr>
<th>Screening Parameters</th>
<th>AF</th>
<th>Cerebral ischemia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean age (years)</td>
<td>60</td>
<td>65</td>
</tr>
<tr>
<td>Cholesterol levels (mg %)</td>
<td>300</td>
<td>300</td>
</tr>
<tr>
<td>Diabetes mellitus (no. of patients)</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>Smoking</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>Hypertension</td>
<td>25%</td>
<td>37.5%</td>
</tr>
</tbody>
</table>

AF = amaurosis fugax.
of clinical atherosclerotic disease in the general population.

The group with amaurosis fugax was remarkably
free of cardiac disease (table 2). None had histories of
angina pectoris, myocardial infarction or valvular dis-
ease or were taking cardiac medications; all had nor-
mal electrocardiograms. In the cerebral ischemic
group, 45% had severe coronary atherosclerosis with
ten patients having documented ST abnormalities or
myocardial infarction. Serum cholesterol levels were
uniformly elevated in both groups.

Both groups were relatively free of diabetes with
one patient of 31 having the adult onset variety. Hypertension was present in both groups. App-
proximately 25% of the patients with amaurosis fugax
had elevated pressures while 37.5% of the cerebral
ischemic group had similar findings. Smoking was
ubiquitous in both groups; 80% of the patients studied
had smoked for 20 or more years. Family history did
not appear helpful in differentiating the two groups of
patients, and a family history for stroke, heart disease,
or severe atherosclerosis obliterans was mixed
between the two groups.

Symptoms of the eight patients with amaurosis
fugax varied from complete loss of vision to partial
field loss, shimmering blank spots, or snow. All of
these complaints were on the ipsilateral side of carotid
stenosis. The remaining 22 patients presented with
symptoms of cerebral ischemia, e.g., transient hemiparesis, loss of sensation or progressive stroke.

Auscultation of the carotid vessels preoperatively
revealed bruits in seven of eight patients (88%) with
AF. An incidence of 41% bruits at the carotid bifurca-
tion in the cerebral ischemic group contrasts with the
previous figure. The presence or intensity of bruits did
not correlate with the degree of stenosis of the carotid
vessels but auscultation of a new bruit or loss of a
bruit showed a high correlation with progression of
the disease process.

Arteriograms of the eight patients with amaurosis fugax showed a minimum stenosis of 75%
of the internal carotid on the symptomatic side (table
3). The carotid bifurcation was always involved and,
in those patients with arch studies, bilaterality of the
disease was the rule. Five of these eight patients had
stenoses of 90% or more of the internal carotid at the
bifurcation, and all had ipsilateral visual symptoms.
One-half of the patients in the cerebral ischemic group
had internal carotid stenosis of greater than 75% with
the range varying from 30% to occlusion on the symp-
tomatic side.

Thirty-four endarterectomies were carried out on
31 patients (table 4). In one instance a vein patch graft
was used in addition to the primary endarterectomy.
The mortality rate was 2.9% with one postoperative
cerebral infarction eight hours after surgery, resulting
in the eventual death of the patient. Morbidity in-
cluded transient hemiparesis in one patient and
myocardial infarction in another. Two patients had
isolated tenth and twelfth nerve deficits postopera-
tively; the latter deficit was permanent. The fifth pa-
tient had a proximal suture line leak requiring re-
operation. On the basis of 34 endarterectomies, the
total morbidity was 15% and the total permanent mor-
bidity was 2.9%.

Pathological specimens removed at surgery
revealed two with ulcerated plaques, three with
organized thrombus, seven with friable fibrinous tags,
and only three with smooth endothelial linings. The
rest of the specimens had rough atheromatous
degeneration.

In review of long-term results, 22 patients were
re-examined. Of the original 31 patients, two were
dead and seven were lost to follow-up. Eight patients
with amaurosis fugax had no recurrence of the
preoperative symptoms when last seen. Fourteen

| TABLE 3 |
| Stenotic Lesions of the Internal Carotid Artery |
| Pr. | Right (%) | Left (%) |
| AF | 95 | 75 |
| EB | 75 | Occlusion |
| NB | 80 | 90 (developed bruit during follow-up) |
| OP | 95 | 90 |
| HC | 80 | 30 |
| HT | 50 | 50 |
| KH | 50 | 50 |
| RT | 95 | 65 |
| SO |  |

CVA = cerebrovascular accident.
cerebral ischemic patients had no recurrence of preoperative symptoms giving a symptomatic cure rate of 100% at the time of re-examination. However, six of the 22 patients (three from the AF group) seen at follow-up were referred for repeat angiography due to progression of disease in the contralateral carotid or basilar systems. Doppler studies revealed progression of disease and loss of flow in the ipsilateral or contralateral internal carotid artery in a number of these patients. Three patients of the 22 examined by Doppler methods (14%) had occlusion of the operated internal carotid artery.

Discussion
Several contrasts appear in these two groups of patients. The visual symptoms of amaurosis fugax were found in the younger group of patients. On questioning the cerebral ischemic patients in follow-up evaluation, a number of them recalled visual spells months to years before onset of motor or sensory deficit. It is our clinical impression that a large number of patients have those visual symptoms and do not seek medical therapy or are not judged seriously ill until transient hemiparesis or sensory loss develops. This study did not yield any differentiating trends between the two groups for hypertension, family history of cardiac disease, stroke, or diabetes.

The patients with amaurosis fugax were free of clinical cardiac disease in comparison with 45% of the cerebral ischemic group who had symptoms of atherosclerotic coronary artery disease. This may have implications for the type of embolism seen in cerebral ischemic patients. It is well documented that patients with calcific heart valves and irregular cardiac rhythms are predisposed to embolization. Calcific cholesterol emboli of cardiac origin are more prone to cause permanent changes in the retina as well as forming occlusive emboli in the cerebral. Thus, severe atherosclerosis involving the heart may predispose to embolization of a more permanent nature with resultant neurological deficits in the form of minor or major strokes. Compare this to patients with amaurosis fugax who are free of motor and sensory deficit, whose symptoms are transient, and whose emboli consist of platelet fibrin aggregates. The data are suggestive but not confirmatory.

Angiographically, patients with amaurosis fugax had a higher degree of internal carotid stenosis while the cerebral ischemic group had a broader range of stenotic lesions of the internal carotid artery. Comparing the two groups, a higher degree of stenosis of internal carotid vessels was found in the amaurosis fugax patients despite only transient visual symptoms. Empirically the severity of stenosis does not parallel the severity of symptoms.

Severe stenosis was not considered the primary indication for operation. Lateralizing signs may occur without significant stenosis. Extrapolating experience from peripheral vascular lesions is misleading, for patients may be just as symptomatic from a 30% stenotic lesion as a 90% stenotic lesion of the internal carotid artery. Sudden stroke may occur without significant stenosis, the only sign of disease progression being the development of a bruit. Currently our indications for operation are lateralizing symptoms of cerebral or retinal ischemia in the setting of significant atherosclerotic disease of the carotid bifurcation. This concurs with other series dealing with AF.

Conclusion
A group of 31 endarterectomy patients were reviewed in an attempt to differentiate patients with amaurosis fugax from those with transient cerebral ischemia. Patients with AF tend to be younger, and had 75% or greater stenosis of the internal carotid artery on the symptomatic side. All of the AF patients had unilateral visual symptoms which cleared with surgery. Of special interest is the fact that these patients were devoid of cardiac disease while 45% of the cerebral ischemic patients had significant myocardial disease. This may reflect the type of transient embolic phenomenon which contributes to amaurosis fugax (i.e., platelet fibrin aggregates). Compare this to calcific cholesterol emboli of cardiac origin which may produce more permanent retinal and cerebral symptoms.

References
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Stroke. 1975;6:493-496
doi: 10.1161/01.STR.6.5.493

Stroke is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0039-2499. Online ISSN: 1524-4628

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