Fibromuscular Disease of Carotid Arteries: Long Term Results of Graduated Internal Dilatation

DAVID S. STARR, M.D., GERALD M. LAWRIE, M.D., AND GEORGE C. MORRIS, JR., M.D.

SUMMARY From 1967 to 1979, 25 patients (pts) were operated on for fibromuscular disease (FMD) of the internal carotid artery (ICA). Eleven patients (44%) had transient weakness of an extremity, 4 had amaurosis fugax and 6 (24%) had an asymptomatic carotid bruit. Bilateral carotid arteriography showed significant stenotic lesions in 23 pts (92%) (bilateral in 10), arterial dissection in 1, and severe associated atherosclerosis in 1. Of these, 22 pts had arteriotomy and graduated internal dilatation (GID) (9 bilateral); 2 underwent GID with ICA endarterectomy and patch graft: 1 pt had tube graft replacement of the ICA.

There was no operative mortality. One pt had a stroke during operation after tube graft replacement of the ICA. Of the 19 pts followed for 2 to 12 years (mean 7.3 years), 2 had late recurrence of mild symptoms. One pt required GID of the contralateral ICA for recurrence of symptoms 4 years postoperatively.

Graduated internal dilatation of fibromuscular disease of the internal carotid artery can produce long term relief of symptoms; recurrence is rare.

FIBROMUSCULAR DISEASE, (FMD) first described in the renal arteries,1 is a unique arteriopathy characterized by areas of fibrous and muscular intimal hyperplasia alternating with aneurysmal dilatations of the arterial wall, giving a "string of beads" appearance on the arteriogram. Since the first description of fibromuscular disease of the internal carotid artery in 1965, different methods of treatment with varying results have been reported. These include resection of the diseased portion of the artery with either end-to-end anastomosis or replacement by autogenous saphenous vein graft,2 excision of fibromuscular tissue and subsequent repair3 and graduated internal dilatation using Bakes dilators4-10 or Fogarty catheters.11 Although short term results appear better using graduated internal dilatation as first described by Morris et al.,1 the long term results of this method are uncertain.

To determine the long term results of the surgical treatment of internal carotid artery FMD by graduated internal dilatation, 25 patients treated by this method over 12 years were followed up.

Patients and Methods
From 1967 to 1979, 25 patients were operated on for fibromuscular disease of the carotid arteries. There were 19 females and 6 males, with an average age of 58 years (fig. 1). Hypertension was present in 4 patients, and was believed to be secondary to fibromuscular disease of the renal arteries in one. Another 4 patients had symptomatic coronary artery disease, and one had aortoiliac atherosclerosis.

Clinical Findings
Symptoms (fig. 1) were similar to those in patients with atherosclerotic carotid artery disease. Eleven patients (44%) had transient ischemic attacks with unilateral transient weakness of the face (3 patients), of the arm (6 patients), or of the arm and leg (2 patients). Only one patient had a stroke in evolution, subsequently found to be due to internal carotid artery occlusion from dissection of asymptomatic FMD. Eighteen patients (72%) reported non-localizing neurological symptoms, including dizziness (10 patients), headache (8 patients), and transient loss of consciousness (2 patients). Four-vessel arteriography failed to reveal a pathological cause for these non-localizing cerebral symptoms.

A loud bruit was the most common physical finding, (17 pts, 68%) and was heard also by the patient in seven of these cases. Hypertension was secondary to renal FMD in one patient, and was subsequently treated by surgery after successful graduated dilatation of the carotid lesion.

Arteriographic Findings
Bilateral carotid arteriograms showed several patterns of disease in these patients, but, predominantly, the classical "string of beads" appearance (20 of 25 patients). There were 4 patients with tubular stenosis, and one with acute dissection. The lumen was significantly (greater than 75%) narrowed in all 25 patients who came to surgery.

The most common area affected was the middle third of the internal carotid artery, while the distal and proximal thirds were usually free of disease. Kinking and tortuosity were common, especially in patients with a normal or high carotid artery bifurcation. The disease was usually bilateral, with significant bilateral lesions in half the patients. There was significant atherosclerosis at the carotid artery bifurcation and in the proximal third of the internal carotid artery in 3 patients.

One patient had bilateral asymptomatic FMD and was followed for 3 years, at which time she developed intermittent lower extremity weakness, especially on standing. Bilateral graduated dilatation relieved these symptoms.

From the Baylor College of Medicine, Houston TX 77030.
GRADUATED DILATATION IN CAROTID FMD/Starr et al. 197

FIGURE 1. Age and sex distribution of 25 patients with fibromuscular disease of the internal carotid arteries.

Technique

Of the 25 patients, 22 had arteriotomy and graduated internal dilatation, (fig. 2) using the method of Morris.9 Nine of these patients required bilateral procedures. (fig. 3). The 3 other patients had severe atherosclerotic lesions proximal to the areas of FMD and required a combination of operative techniques. Two patients had carotid endarterectomy followed by graduated internal dilatation and Dacron patch angioplasty, while a third, who had a totally occluded internal carotid artery due to dissection of a fibromuscular area distal to an atherosclerotic plaque, also required replacement of the internal carotid artery to within 0.5 cm of the base of the skull with a Dacron tube graft (occlusion time 31 min).

The operative technique of graduated internal dilatation has been described in detail previously.9 Briefly, all patients were operated on under general endotracheal anesthesia. In 24 patients the internal carotid artery was approached anterior to the sternomastoid, while in one patient the posterior approach was used to expose the internal carotid artery at the base of the skull.10

Exposure of the internal, external and common carotid arteries was achieved as for a carotid endarterectomy, ensuring that the diseased section of the internal carotid artery was visualized, and that the artery could be straightened out between the bifurcation and the base of the skull by gentle traction. This sometimes involved mobilizing the internal carotid artery to the level of the styloid process, and dividing the carotid sinus nerve.

Once the internal, external and common carotid arteries were exposed, they were clamped, and then a transverse arteriotomy was carried out in the proximal third of the internal carotid artery below the level of the disease. Before dilatation was commenced, downward traction was exerted on the artery to ensure that it lay straight between the arteriotomy and the base of the skull, and that visualization of the diseased section was achieved. Bile duct (Bakes) dilators were then passed anterograde up the internal carotid artery, commencing with a 3 or 3.5 mm size and proceeding to 5 or 5.5 mm in 0.5 mm increments (fig. 2). Heparin was not used unless a more involved procedure than simple dilatation was required, in which case 1000 units of heparin in 10 cc of saline were introduced into the internal carotid artery after arteriotomy, and the clamp was then replaced. Inadvertent distal embolization of air by the “plunger” mechanism was avoided by allowing adequate backflow of blood from the internal carotid artery by release of the appropriate clamp before introduction of the dilator. During initial passage of the dilator distally, the membranes associated with FMD could be felt giving way until the dilator stopped at the base of the skull. Then dilatation felt smoother as the artery gradually expanded.

The transverse arteriotomy was then closed with interrupted 6/0 polypropylene sutures and the clamps removed after the area was flushed.

FIGURE 2. Technique of graduated internal dilatation. 2a) Transverse arteriotomy performed at the origin of the internal carotid artery. 2b) Tortuous internal carotid artery is straightened by traction. 2c) Membranes and stenotic areas are dilated by gentle passage of Bakes dilators. 2d) Transverse arteriotomy closed with interrupted polypropylene.
TABLE 1  Symptoms at Presentation in 25 Patients

<table>
<thead>
<tr>
<th>Symptom</th>
<th>Count</th>
</tr>
</thead>
<tbody>
<tr>
<td>Transient ischemic attacks</td>
<td>11</td>
</tr>
<tr>
<td>Amaurosis fugax</td>
<td>4</td>
</tr>
<tr>
<td>Asymptomatic bruit</td>
<td>6</td>
</tr>
<tr>
<td>Mental slowness</td>
<td>3</td>
</tr>
<tr>
<td>Neck pain</td>
<td>1</td>
</tr>
<tr>
<td>Non-localizing neurological symptoms</td>
<td>18</td>
</tr>
</tbody>
</table>

Mean occlusion time of the internal carotid was 11.3 min (range 7.0 to 31.0 min).

Follow Up

Nineteen of the 25 patients (19/25), were followed up, either by readmission and arteriography (5/19), outpatient examination (5/19), examination by the patient's local physician (3/19), or follow up by questionnaire and telephone (3/19). Mean follow up period was 7.3 years (range 1 to 12 years).

Results

There was no operative mortality. One patient had a stroke after Dacron tube graft replacement of a severely atherosclerotic internal carotid artery, and even after therapy remained severely disabled from a hemiparesis.

Seventeen of the 19 patients had no symptoms of cerebrovascular disease at follow up. Two patients with preoperative symptoms of dizziness and transient ischemic attacks reported intermittent dizziness at follow up (9 years and 12 years) but did not wish to be reinvestigated at that time. Another patient originally had dilatation of the left internal carotid for amaurosis fugax, and 4 years later had dilatation of the contralateral internal carotid for similar symptoms on the other side, resulting in relief of symptoms.

However, the other 9 patients with bilateral disease had a similar degree of severity on both sides and had bilateral operations.

Follow up carotid arteriograms were made in 5 patients and demonstrated patency of the internal carotid from 1 to 8 years after dilatation (mean 4.5 years). Three of these patients had persistence of preoperative tortuosity of the internal carotid artery, and in 2 there was intimal irregularity suggestive of persistent or recurrent disease. Two other patients had no radiological evidence of FMD.

Of the 14 patients who did not have follow up arteriograms, 4 had oculoplethysmography during the follow up period. Three patients had normal intraocular pressures and remained asymptomatic, findings confirmed by ophthalmodynamometry.

The 19 symptomatic patients had good relief after this operation and these results persisted in the long term. Only 2 patients had mild recurrence of preoperative symptoms (fig. 3).

Discussion

Etiology

Fibromuscular dysplasia of the internal carotid artery is an uncommon disease, with an incidence in one series of 21 patients in 7000 who had carotid arteriography. It occurs predominantly in middle aged women, and this observation, when combined with the known changes in the media of arteries in pregnancy has led to speculation of a hormonal etiology. There must, however, be other factors to account for its predominant occurrence in the renal arteries, and less commonly, in the internal carotid, vertebral and external iliac arteries. Stanley has proposed a mechanical theory of causation based upon proposed unusual stresses acting on a larger than normal artery at an area poorly supplied by vasa vasorum. Given a suitable hormonal milieu, the media is damaged, and disruption of the internal elastic lamina followed by attempted repair then leads to one of the histological types of FMD. Despite a variety of proposed explanations, the etiology still remains unknown.

Pathology

The histological appearance of FMD, first described by Wylie and Wellington, typically consists of medial thickening due to hypertrophy of the smooth muscle and fibrous elements. Disruption of the internal elastic lamina occurs adjacent to these hypertrophic segments of diseased muscle. Intimal involvement is usually minimal and confined to localized cellular proliferations, but these may cause circumferential "membranes" to project out into the lumen. Arteriosclerosis may also be present. Gross appearance varies from early hyperplastic changes to marked beading and tortuosity with multiple stenotic areas.

Angiographic Appearance

Although the usual angiographic appearance of FMD is the "string of beads," a recent review describes the tubular and localized forms in addition to the classical type. In addition, one patient in this series with dissection, had an angiographic finding which Effeney et al. suggests should stimulate a search for FMD elsewhere in the body. Spontaneous dissection of areas of FMD can occur in an otherwise asymptomatic patient and the possibility of this should prompt the physician to consider operation in patients who are asymptomatic but have definite FMD upon carotid arteriography. The only morbidity in this series was a postoperative stroke in a patient who had a totally occluded internal carotid artery, and at operation was found to have dissection of an area of
FMD. A tube graft was used to replace the internal carotid artery. In view of the possible catastrophic effects of sudden dissection and total occlusion of an internal carotid artery affected by FMD it would seem reasonable to advocate prophylactic operation even in asymptomatic patients, especially if the angiographic appearance suggested marked change. However, there are insufficient data properly to analyze this position statistically.

Symptoms

The origin of symptoms in these patients is unclear. Possibilities include embolization of clot or platelet aggregates from the aneurysmal areas, or cerebral hypoperfusion due to one or multiple areas of critical stenosis. In view of the pathology, where "membranes" may project out into the lumen, the second of these possibilities may seem more likely, but often the clinical presentation may suggest embolism, as in the case of amaurosis fugax.

Despite the pervasive belief that this is a benign disease in which progression or symptoms are rare, FMD can progress and cause symptoms, and even proceed to occlusion.19,21 One asymptomatic patient in this series was followed for 3 years at which point symptoms appeared. She then had surgical correction with a good result. Symptoms in the other patients were often incapacitating, and treatment allowed return to a normal life. Although only patients in this series presented with a stroke in evolution, experience of other workers19,21-23 suggest that stroke complicates this condition in about 20% of the reported patients. This suggests a need to consider intervention in symptomatic patients.

Choice of Procedure

Because the dysplasia commonly involves the middle third of the extracranial internal carotid artery, the methods of arteriotomy and vein patch, or resection and vein interposition, are not generally applicable, as these techniques require exposure of the internal carotid artery distal to the disease. For this reason, the technique of graduated internal dilatation, as described by Morris et al.,8 seems to be the best method for most patients. However, the internal carotid can be exposed at the base of the skull, if necessary, by the approach which is posterior to the sternomastoid.12

Dilatation of stenotic arterial lesions using low compliance balloon catheters (percutaneous transluminal angioplasty)24 has recently been popular. This technique has been particularly successful in fibromuscular dysplasia of the renal arteries,25 and this naturally suggests its use in FMD of the carotid arteries. There are no reports at present of the use of this technique in the carotid arteries. The recognized incidence of distal embolization during balloon dilatation,24 and the tortuosity of the carotid artery commonly occurring in FMD may limit its usefulness in this situation.

References

Fibromuscular disease of carotid arteries: long term results of graduated internal dilatation.
D S Starr, G M Lawrie and G C Morris, Jr

doi: 10.1161/01.STR.12.2.196

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://stroke.ahajournals.org/content/12/2/196

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Stroke can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Stroke is online at:
http://stroke.ahajournals.org/subscriptions/