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AJNR Am J Neuroradiol 1981, 2 (2) 175-180

<http://www.ajnr.org/content/2/2/175>

This information is current as of May 11, 2024.

Fibromuscular Dysplasia of the Internal Carotid Artery: Percutaneous Transluminal Angioplasty

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Percutaneous transluminal angioplasty was used to successfully dilate critical narrowings of the internal carotid artery caused by fibromuscular dysplasia. The technique of this new procedure is presented along with the angiographic findings and results in three patients. Angioplasty may be used as an alternative to open arteriotomy and graduated dilatation in treating stenosis due to symptomatic fibromuscular dysplasia in the immediate extracranial part of the internal carotid artery.

Fibromuscular dysplasia as an angiographic diagnosis is made most often in the renal arteries where 85% of cases are found [1]. The second most common location is in the internal carotid artery where it constitutes the second most common cause for extracranial carotid narrowing [2, 3]. About 1% of all carotid angiograms show fibromuscular dysplasia [2]. Usually the C1-C2 part of the internal carotid artery is involved and frequently (60%-65%) the involvement is bilateral [2, 4]. About 50% of patients have associated renal arterial lesions diagnostic of fibromuscular dysplasia [4, 5]. While true intracranial fibromuscular dysplasia is rare, affected patients also have an increased incidence of intracranial aneurysms and intracranial neoplasms [2, 4, 6].

The etiology is unclear. The disease is sometimes progressive [4, 7, 8] and is often associated with significant clinical findings, such as transient neurologic deficits, completed strokes, or subarachnoid hemorrhage [2, 4, 5, 9, 10]. Surgical treatment has commonly been endarterectomy, excision of the involved segment, or graduated internal dilatation [1, 5, 9, 10]. The latter procedure uses bile duct dilators inserted through open arteriotomy in the distal common carotid or proximal internal carotid artery and then passed through the stenotic segments.

Recently there has been increasing interest in using percutaneous transluminal angioplasty to treat atherosclerotic stenoses. This interest has heightened since 1974 when Grüntzig introduced a double-lumen balloon dilating catheter [11]. Recent reports have described the successful use of this catheter in the treatment of iliac, femoral, popliteal, renal, and coronary artery stenoses [12-17]. To our knowledge, there has been no reported treatment of fibromuscular dysplasia of the internal carotid artery by percutaneous transluminal angioplasty. Percutaneous transluminal angioplasty has been used for dilating an internal carotid artery stenotic lesion of unknown origin [18].

Case Reports

Three patients admitted to the Loma Linda University Medical Center had complete angiographic documentation of the cervicocranial circulation.

Received April 14, 1980; accepted after revision October 29, 1980.

Presented at the annual meeting of the American Society of Neuroradiology, Los Angeles, March 1980.

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This article appears in March/April 1981 *AJNR* and May 1981 *AJR*.

AJNR 2:175-180, March/April 1981
 0195-6108/81/0022-0175 \$00.00
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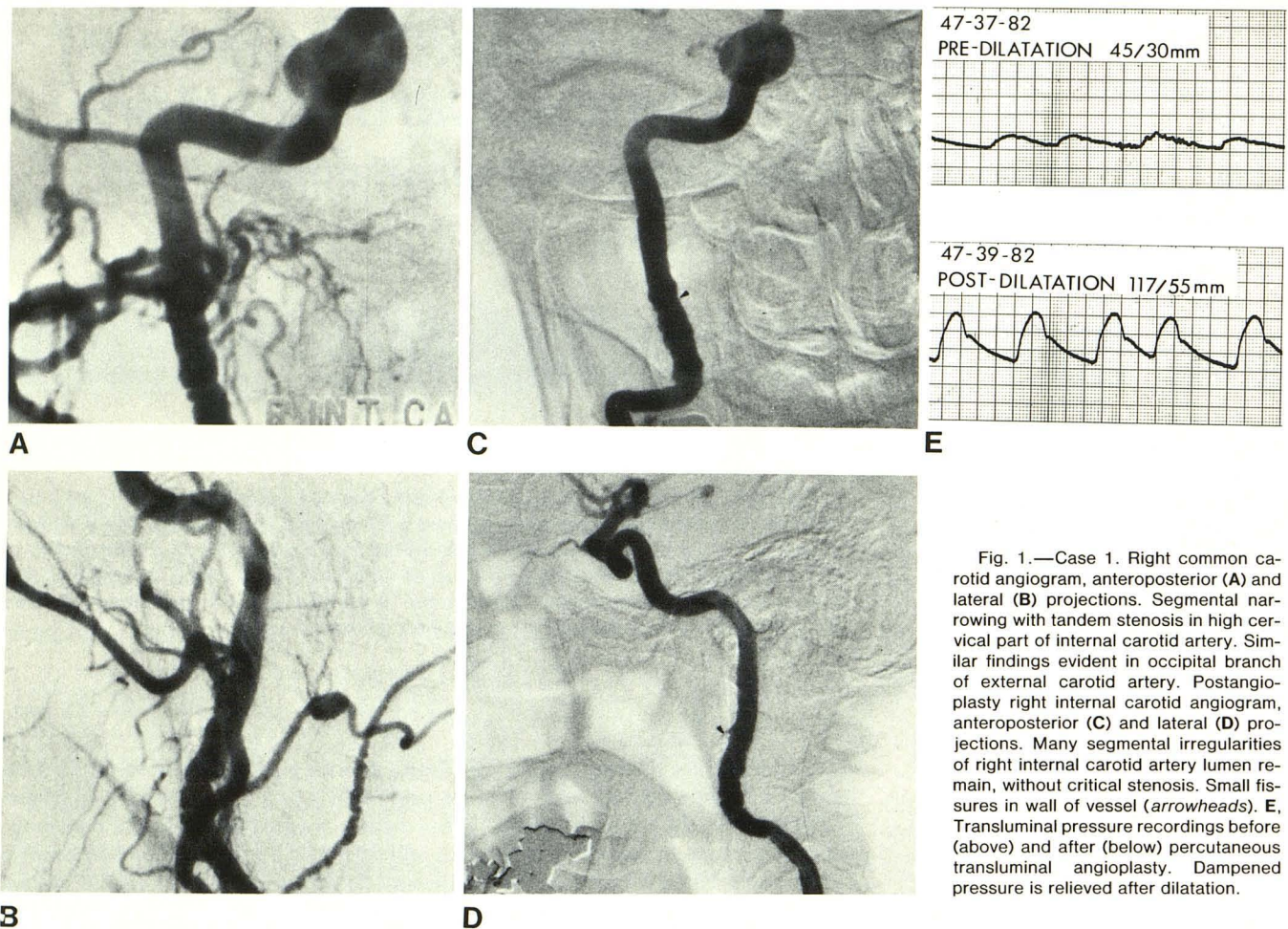


Fig. 1.—Case 1. Right common carotid angiogram, anteroposterior (A) and lateral (B) projections. Segmental narrowing with tandem stenosis in high cervical part of internal carotid artery. Similar findings evident in occipital branch of external carotid artery. Postangioplasty right internal carotid angiogram, anteroposterior (C) and lateral (D) projections. Many segmental irregularities of right internal carotid artery lumen remain, without critical stenosis. Small fissures in wall of vessel (arrowheads). E, Transluminal pressure recordings before (above) and after (below) percutaneous transluminal angioplasty. Dampened pressure is relieved after dilatation.

Case 1

A 74-year-old woman had the classic "string of beads" appearance of fibromuscular dysplasia with a prominent stenosis among several tandem segmental narrowings in her right internal carotid artery (figs. 1A and 1B). Similar findings were evident in the left internal carotid artery, but without significant stenosis. The vertebral arteries were not involved. Several days later a 7 French Grüntzig balloon dilating catheter (Cook) was introduced transfemorally over a 260-cm-long exchange guide wire which had been selectively placed through a conventional catheter into the right common carotid artery. After hyperextension of the neck to straighten a vascular kink, the balloon catheter was carefully placed into the stenotic part of the internal carotid artery with a "floppy" guide wire leader. A pressure record showed decreased pressure distal to the stenosis (fig. 1E). The balloon was inflated with dilute contrast material for 5–10 sec, then deflated, and another pressure record was obtained showing improved pressures (fig. 1E). The collapsed balloon catheter was withdrawn; postdilatation internal carotid angiogram showed obliteration of the stenosis (figs. 1C and 1D). There was a small fissure in the wall of the vessel probably due to a "fracture" secondary to the dilatation. The patient tolerated the procedure well and had relief of transient dizziness and light-headedness which had been her most significant complaints before the procedure. The patient has been followed at 3 monthly intervals and when last seen she had resumed full activities at home. There were no neurological deficits and no carotid bruits.

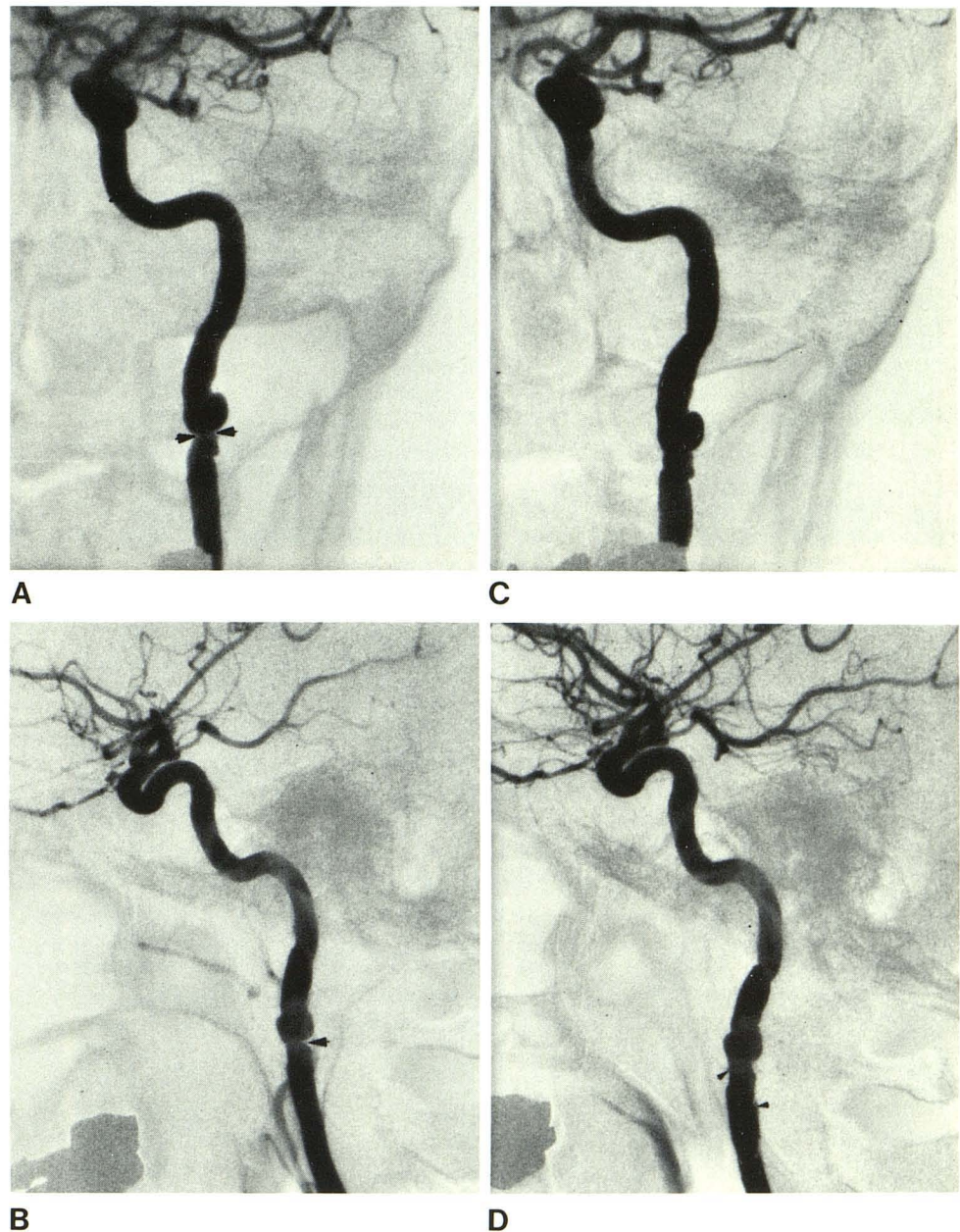
Case 2

A 53-year-old woman had a noncircumferential weblike narrowing just proximal to a diverticulum in the wall of her left internal carotid artery (figs. 2A and 2B). The area of greatest stenosis was dilated in a similar manner to case 1. No pressure recordings were obtained due to technical problems. After the procedure she had relief of her major complaint of pulsatile ringing in the left ear, aggravated by exercise. The tinnitus had been of sufficient magnitude to cause sleeplessness. She also had a left neck bruit which disappeared after the procedure. Ophthalmodynamometry of the left retinal artery was 90/40 mm before dilatation and 108/45 mm after dilatation. The postdilatation internal carotid angiogram showed less stenosis with a small fissure in the wall of the artery, again thought to represent a "fracture" secondary to the dilatation procedure (figs. 2C and 2D). At the 1 and 3 month follow-up examinations, she continued to be neurologically intact and had no audible cervical bruits.

Case 3

A 53-year-old woman had severe proximal stenosis and segmental narrowing of the extracranial part of her left internal carotid artery (figs. 3A and 3D). The stenosis was dilated with a Grüntzig 7 French balloon catheter with documentation of improved pressure recordings after the procedure in comparison to before the procedure (fig. 3G). The immediate postdilatation angiogram showed two

Fig. 2.—Case 2. Left internal carotid angiogram, anteroposterior (A) and lateral (B) projections. Critical noncircumferential stenosis in high cervical part of left internal carotid artery just proximal to eccentric diverticulum in wall of vessel (arrowheads). Postangioplasty left internal carotid angiogram, anteroposterior (C) and lateral (D) projections. Stenosis has been significantly relieved. Small fissures in wall of vessel (arrowheads).



small fissures in the posteromedial wall of the left internal carotid artery (figs. 3B and 3E).

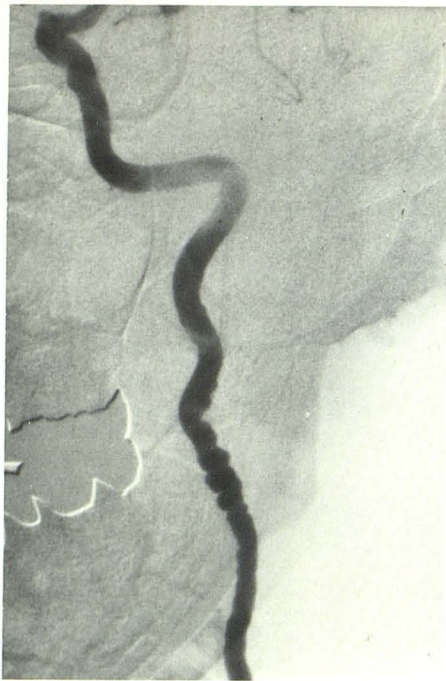
The patient had had a 2 year history of transient ischemic attacks characterized by heaviness of the right upper extremity, weakness of the right lower extremity, and amaurosis fugax in the left eye. There were occasional episodes of verbal aphasia. She had a minor stroke about 3 months before angioplasty with mild residual right-sided weakness. Just before the angioplasty, she had recurrent transient ischemic attacks as often as two to three times a day.

The episodes decreased in severity and frequency after the angioplasty. In a 6 week period she had a total of three episodes of right arm and/or leg weakness. One episode was associated with "blurring out" of vision in the left eye. Repeat angiography to rule out residual or recurrent stenosis showed a focal outpouching at the site of a previous small "fracture" without evidence of a flow limiting lesion (figs. 3C and 3F).

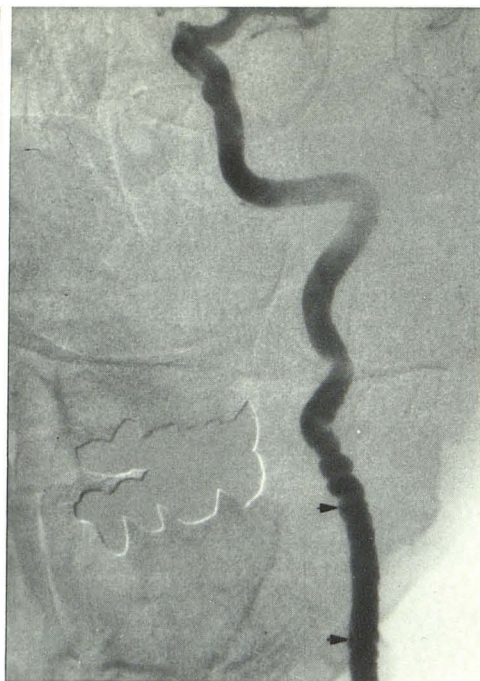
At the time of follow-up angiography, she had a normal-appearing left ocular fundus with a sharp disc and no hemorrhages, exudates, evidence of blocked vessels, or retinal edema. When seen 1 month later, she complained only of heaviness in the right arm and leg which related to the minor stroke she has sustained 3 months before the angioplasty. It is significant that she felt so much better that she requested an angioplasty to be performed on the disease in her right internal carotid artery. This was planned at some further date if she became neurologically symptomatic.

Comment

The patients had the procedure performed while awake and without systemic heparinization. Complete anticoagulation during the procedure was believed to be contraindicated, in that an intimal tear leading to massive dissection or an intramural hematoma would



A



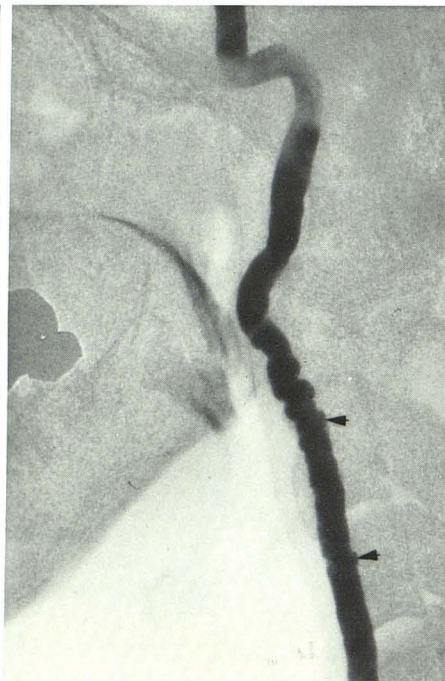
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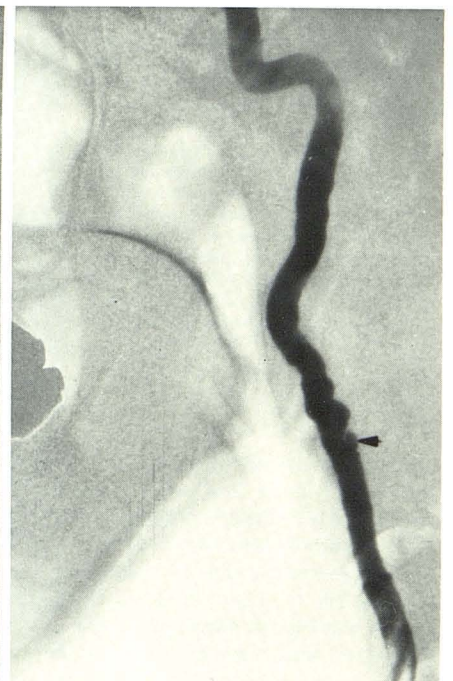
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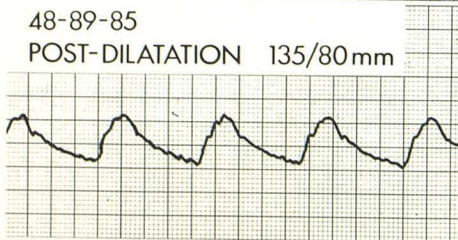
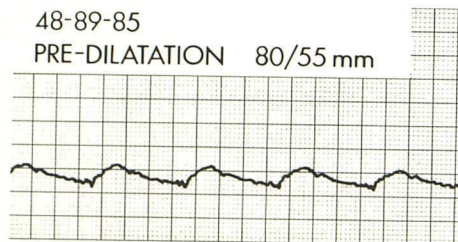
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F



G

Fig. 3.—Case 3. Anteroposterior left internal carotid artery angiograms before (A), immediately after (B), and 6 weeks after (C) percutaneous angioplasty. A, Long diseased segment from C3 to C1 characterized proximally by critical stenosis. B, Two small fissures (*arrowheads*) resulting from angioplasty. Improved flow through narrowed segment. C, Healing of inferior fissure, but continued evidence of an intimal dissection distally (*arrowhead*). Lateral left internal carotid angiograms before (D), immediately after (E), and 6 weeks after (F) percutaneous angioplasty. Greatest stenosis is relieved after angioplasty. E, Two tiny fissures in wall of artery (*arrowheads*). F, Residual intimal lesion (*arrowhead*). No recurrent stenosis. G, Transluminal pressure recordings before (above) and after (below) percutaneous transluminal angioplasty. Improved pulse pressure with more normal wave form after dilatation.

be aggravated if the patients were heparinized. However, the patients were placed on oral aspirin therapy for about 72 hr before and after the procedure.

Discussion

Angiographically, fibromuscular dysplasia is manifested as a "string of beads" (80%–85% of cases), long segment tubular stenosis (6%–12% of cases), or ovoid outpouchings with noncircumferential weblike narrowings (4–6% of cases) [2, 4]. Pathologically, the "string of beads" appearance is usually caused by medial fibroplasia, while tubular stenosis or outpouchings with webs may be caused by any histologic type of fibromuscular dysplasia [2, 4, 19].

Surgical treatment has been resection or endarterectomy in surgically accessible lesions involving the proximal part of the internal carotid artery. When the process involves the distal part of the extracranial internal carotid artery, graduated internal dilatation through open arteriotomy has been the procedure of choice.

Percutaneous transluminal angioplasty probably offers certain advantages to surgical dilatation in the treatment of stenosis of the internal carotid artery caused by symptomatic fibromuscular dysplasia. These advantages include a technically simple procedure performed in an awake patient under local anesthesia. The upper limits of the stenosis can be identified fluoroscopically, avoiding injury to the vessel within the carotid canal. Postdilatation angiography is facilitated by using the same balloon catheter and puncture site in the groin. There are no sutures involved and there is a smooth transition in the lumen of the vessel, decreasing turbulent flow and possible subsequent thrombus formation.

Certain precautions need to be maintained during the performance of angioplasty. The balloon is repeatedly filled with dilute contrast material and emptied upside down to remove any trapped air bubbles before inserting it in the patient. During the dilatation procedure, the patient's neck must be hyperextended to minimize wedging of the balloon catheter.

The stenosis of fibromuscular dysplasia is easily relieved [20] and overdistension of the balloon must be avoided by monitoring the degree of inflation (fig. 4). This is done by using a pressure manometer attached to the balloon catheter and not exceeding the listed maximum pressures recommended by the manufacturer. At no time should the balloon catheter be advanced without a "floppy" guide wire leader. It has been shown that both spontaneous and iatrogenic dissection is associated with fibromuscular dysplasia and all attempts should be made to avoid this complication [6, 7, 21]. The balloon catheter should be left in the stenotic segment for less than 30 sec with continuous pressure transducer recordings through a manifold system. Significant spasm must be avoided by careful slow movements of catheters and guide wires. Since potential complications include thrombosis or embolization of material into the ophthalmic and/or intracranial circulation, patients should be followed by clinical colleagues along with the radiologist.

The production of fissures in the arterial wall is not surprising, since the same finding has been seen in cases surgically dilated by bile duct dilators [5, 9]. Experimentally,

Fig. 4.—Double-lumen balloon catheter in left internal carotid artery. Dilute contrast outlines size and shape of balloon. Location of balloon and tip can be documented fluoroscopically during procedure.



it has been shown that the fissures in the vessel walls after angioplasty are caused by splitting both the intima and media which leads to an increase in the luminal diameter [22, 23]. In fibromuscular dysplasia these splits appear as fissures in the vessel walls.

It is interesting to speculate whether there is durable lumen patency. Some cases of fibromuscular dysplasia treated by open graduated internal dilatation have remained widely patent on follow-up angiography several years later [9]. Long-term control of hypertension after fibromuscular dysplasia angioplasty in the renal arteries has also been reported [24]. In case 3 we were able to demonstrate patency 6 weeks after angioplasty (figs. 3C and 3F). Additional long-term follow-up is being planned.

ACKNOWLEDGMENTS

We thank James Simmons for photographic assistance and Sheila Wills for secretarial assistance.

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