

Sténoses non athéroscléreuses des artères rénales

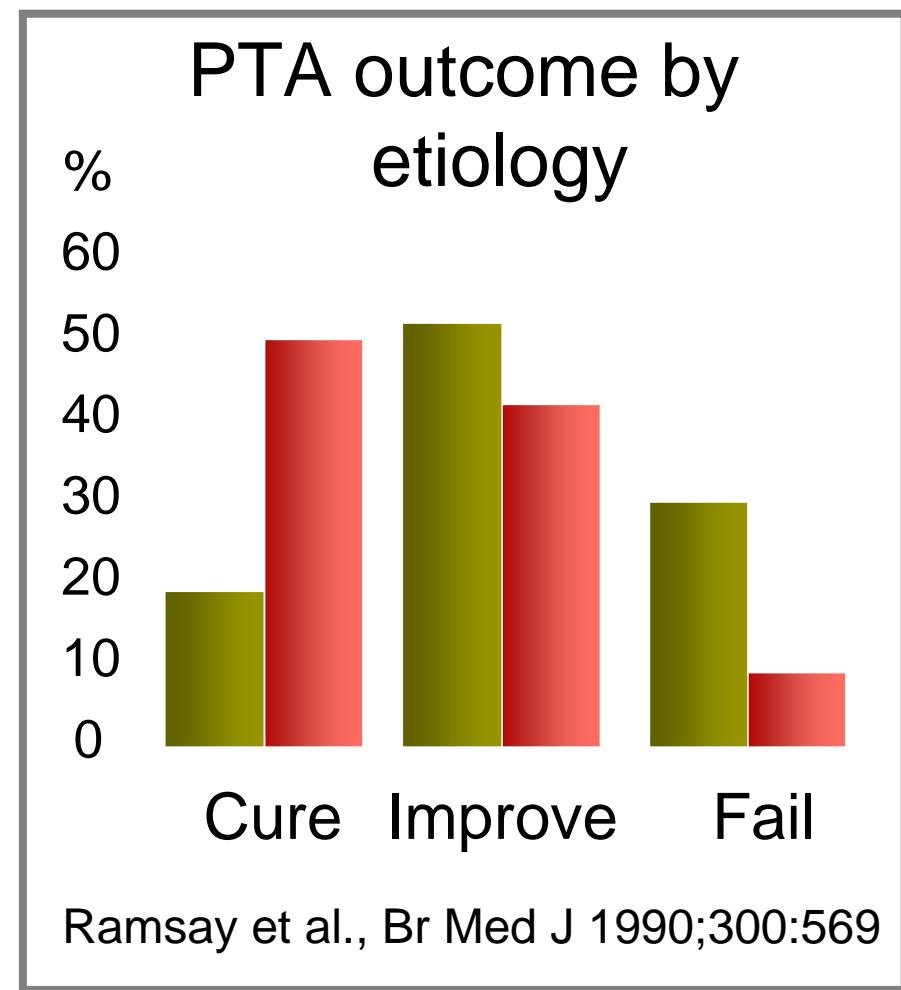
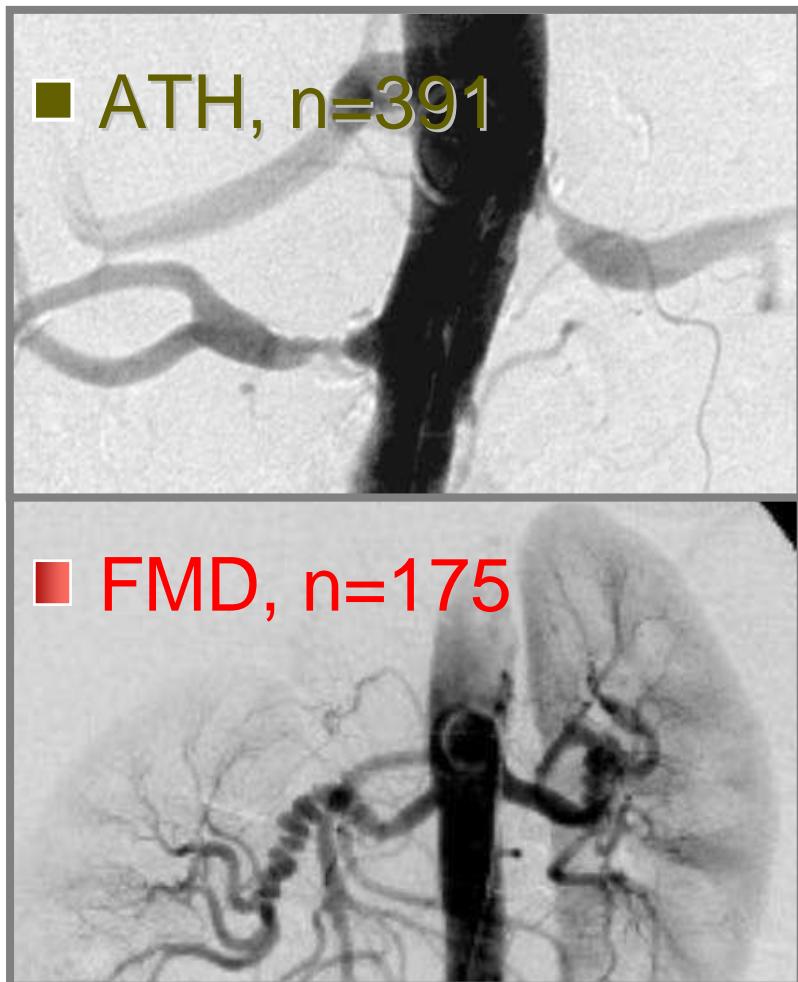


Centre de référence des
maladies vasculaires rares

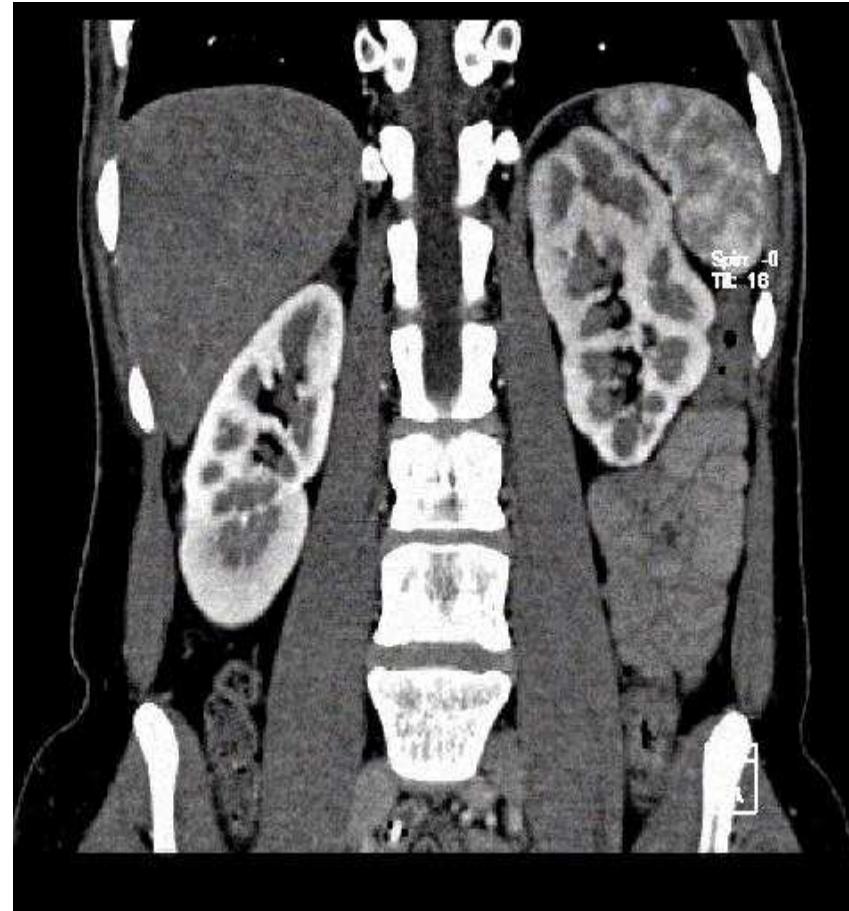
PF Plouin, Unité d'Hypertension
et centre de référence des
maladies vasculaires rares, HEGP

Renal artery stenosis

atherosclerosis v fibromuscular dysplasia



Angio-scanner chez une femme de 38 ans



Pathologic classification

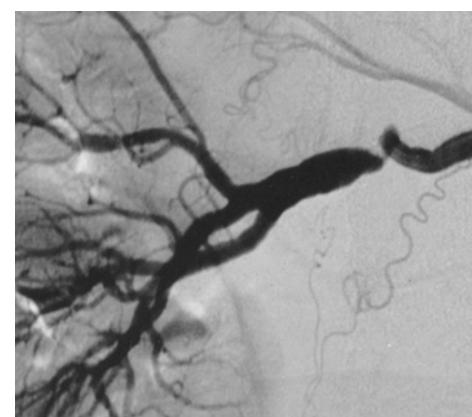
- Based on the dominant wall layer involved¹
 - **Intimal (~5%)** Mesenchymal cells in a loose matrix of subendothelial connective tissue, fragmented internal elastic lamina
 - **Medial (~85%)** Collar of elastic tissue that present as multiple stenoses and intervening aneurismal outpouchings, preserved internal elastic lamina
 - **Perimedial (~10%)** Excessive tissue deposition at the junction of the media and adventitia
- The three types are not mutually exclusive^{1,2}

¹ Stanley JC *In* Renal Vascular disease. WB Saunders, London 1996

² Alimi Y et al, Ann Vasc Surg 1992;6:220

Pathologic-angiographic correlations

	String of beads	Focal stenosis	Tubular stenosis
Intimal, %	0	13	2
Medial, %	66	8	9
Periarterial, %	0	0	1



Most suggestive,
all medial

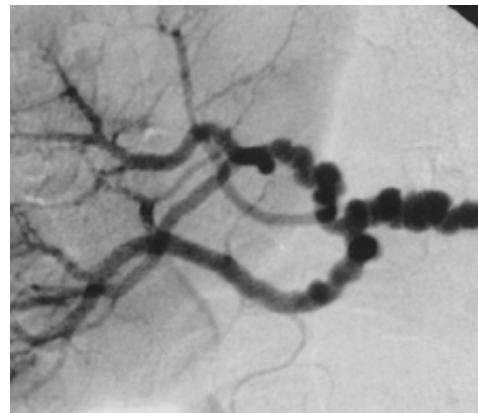
Differ by stenosis length
with an arbitrary cut-off, diverse

¹ Kincaid OW et al, Am J Roentgenol 1968,104:271 (n=60)

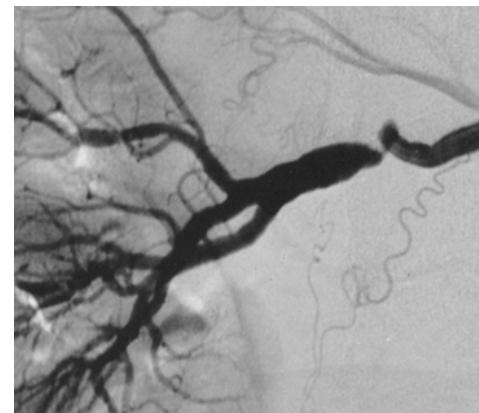
² McCormack LJ et al, Am Heart J 1966,72:188 (n=67)

Arterial phenotypes

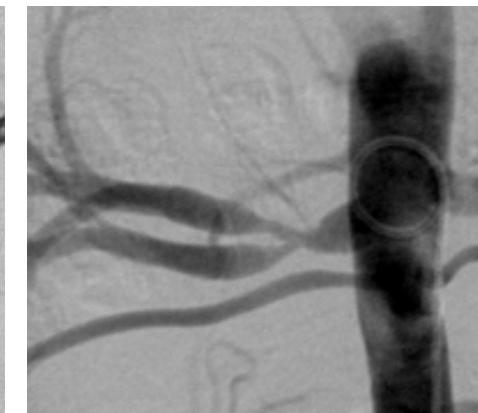
String
of beads



Focal
stenosis



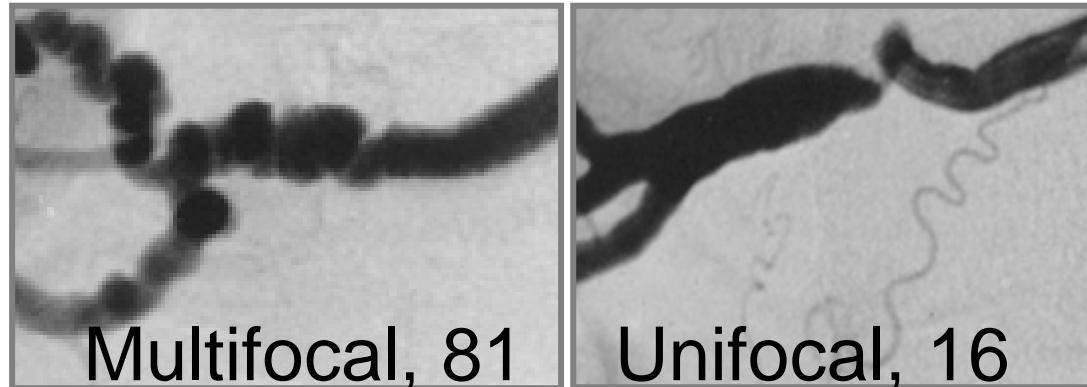
Tubular
stenosis



Multifocal,
medial FMD

Unifocal,
undertermined FMD

Clinical presentation by arterial phenotype



Female	76 (94%)	11 (69%)
Age, y	47 ± 12	28 ± 9
Bilateral	45 (55%)	2 (12%)
Stenosis >75%	37 (45%)	15 (94%)
Small kidney	11 (13%)	9 (56%)

p<0.01 for all
comparisons

Pannier-Moreau I et al, J Hypertens 1997,15:1797

Etiologic factors in renal FMD

- The female predominance suggests a role of estrogens
- Smoking more prevalent in FMD than in matched controls

	FMD	controls	p
Gravidity	3 ± 2	3 ± 3	0.61
OC users, %	55	54	0.77
Ever smoked, %	79	53	<0.01
Kidney mobility, mm	55 ± 46	41 ± 32	0.24
HLA DRw6, %	43	20	0.07

Sang CN & al, Hypertension 1989;14:472
Controls: normotensive renal transplant donors

Outcome of angioplasty in renal artery DFM

Series with ≥ 30 patients followed-up after PTA

	No	Technical success %	HTN cure %	Complic %
Sos, 1983	31	87	59	6
Jeunemaitre, 1989	34	83	65	11
Tegtmeier, 1991	66	100	39	13
Bonelli, 1995	105	89	22	11
Jensen, 1995	30	97	39	3
Klow, 1998	49	98	26	0
De Fraissinette, 2003	70	94	14	11
Alhadad, 2005	59	95	24	
Overall	444	91.6	32.2	9.4

Why BP outcome may be disappointing?

- Hypertension not related to FMD
 - FMD lesions frequent in normotensive subjects
 - Hypertensive patients may have FMD without critical stenosis
- Renal revascularization not obtained
 - Technical failure/restenosis
 - Bilateral lesions/systemic disease

Asymptomatic renal artery FMD

	Angiographic criteria	n FMD/ n donors	%
Spring	NR	14/444	3.2
Cragg	NR	71/1862	3.8
Neymark	'beaded mural irregularity'	47/716	6.6
Andreoni	NR	7/159	4.4
Overall		139/3181	4.4%

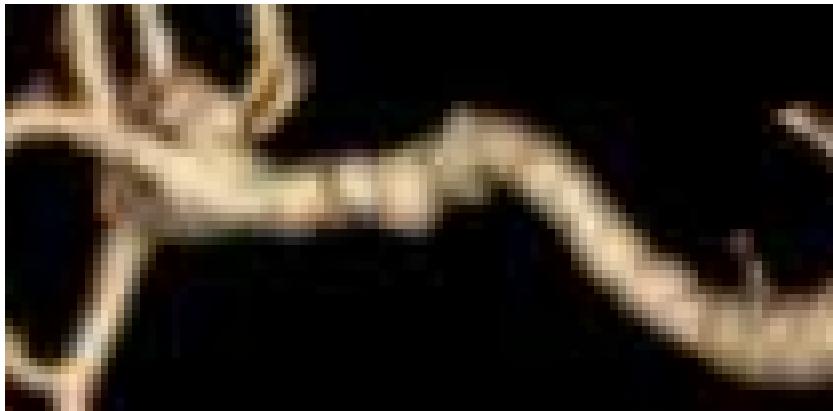
Spring DB, Radiology 1979;133:45. Cragg AH, Radiology 1989; 172:145 Neymark E, Radiology 2000;214:755. Andreoni KA, Transplantation 2002;73:1112

The prevalence of clinically significant renal artery FMD is estimated to be about 0.4%

Plouin PF et al, Orphanet J Rare Dis 2007;2:28

Stenosis quantification in FMD

Notoriously difficult in multifocal FMD



Indirect approaches

Number of diaphragms

Combination of grade and length

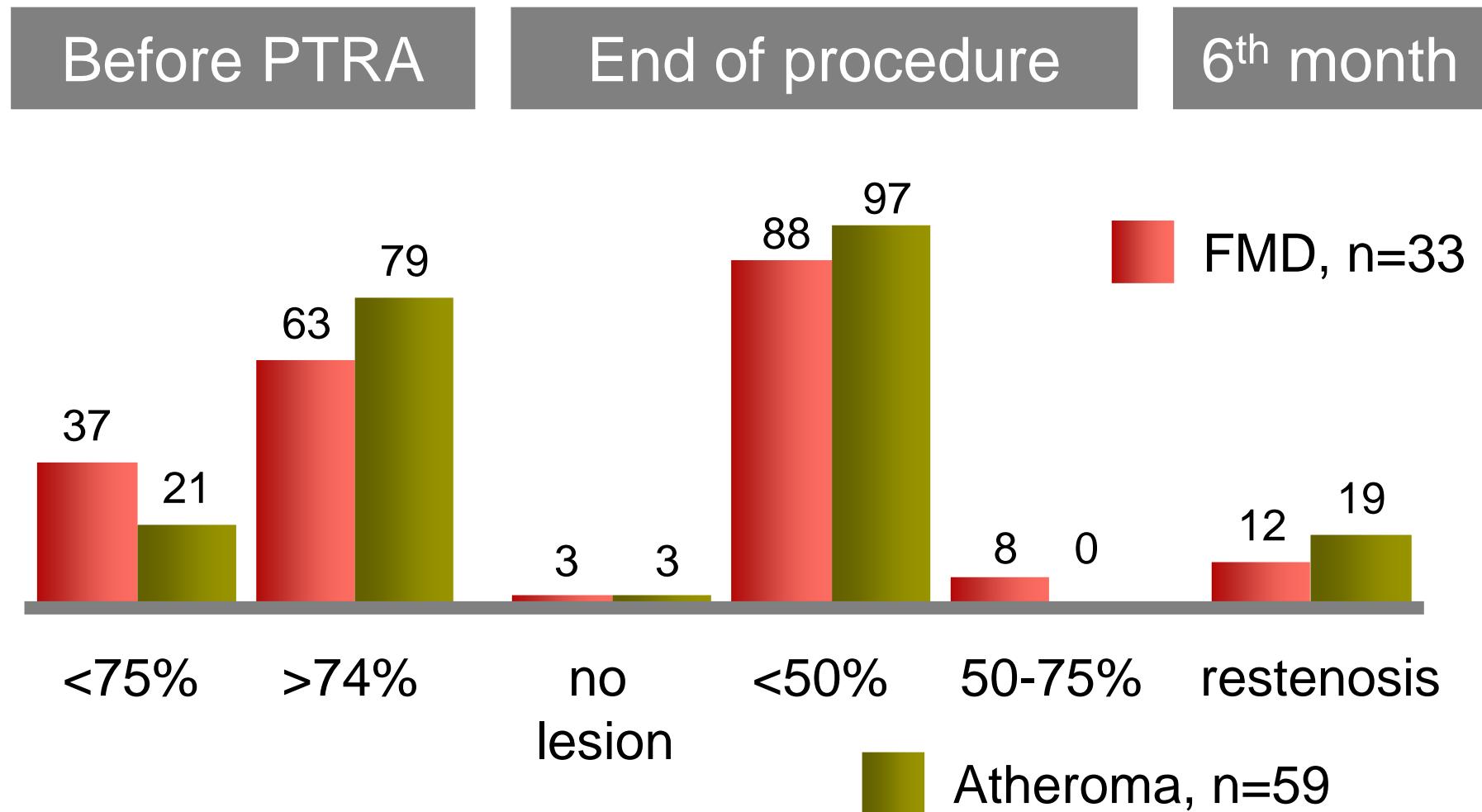
Transstenotic gradient

Collateral circulation

Gold standard?



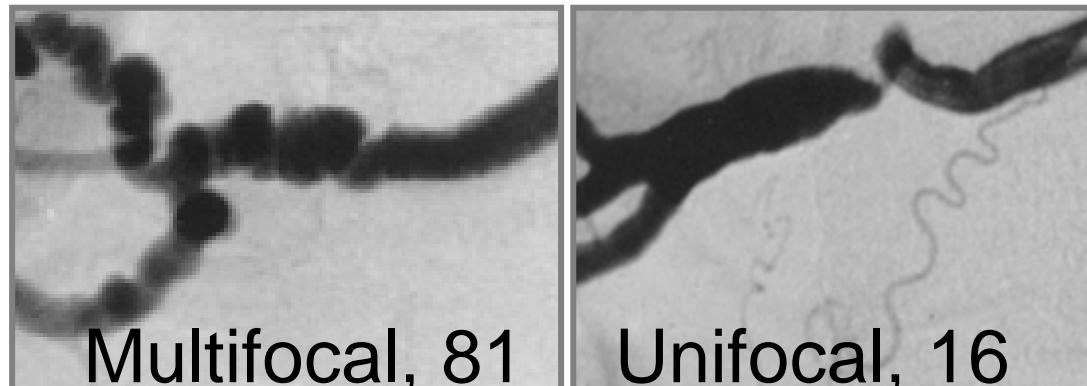
Angiographic outcome in atheroma and FMD



Plouin PF et al., Hypertension 1993;21:89

DSA blinded, coded and read in a random order by two radiologists

Presentation by arterial phenotype

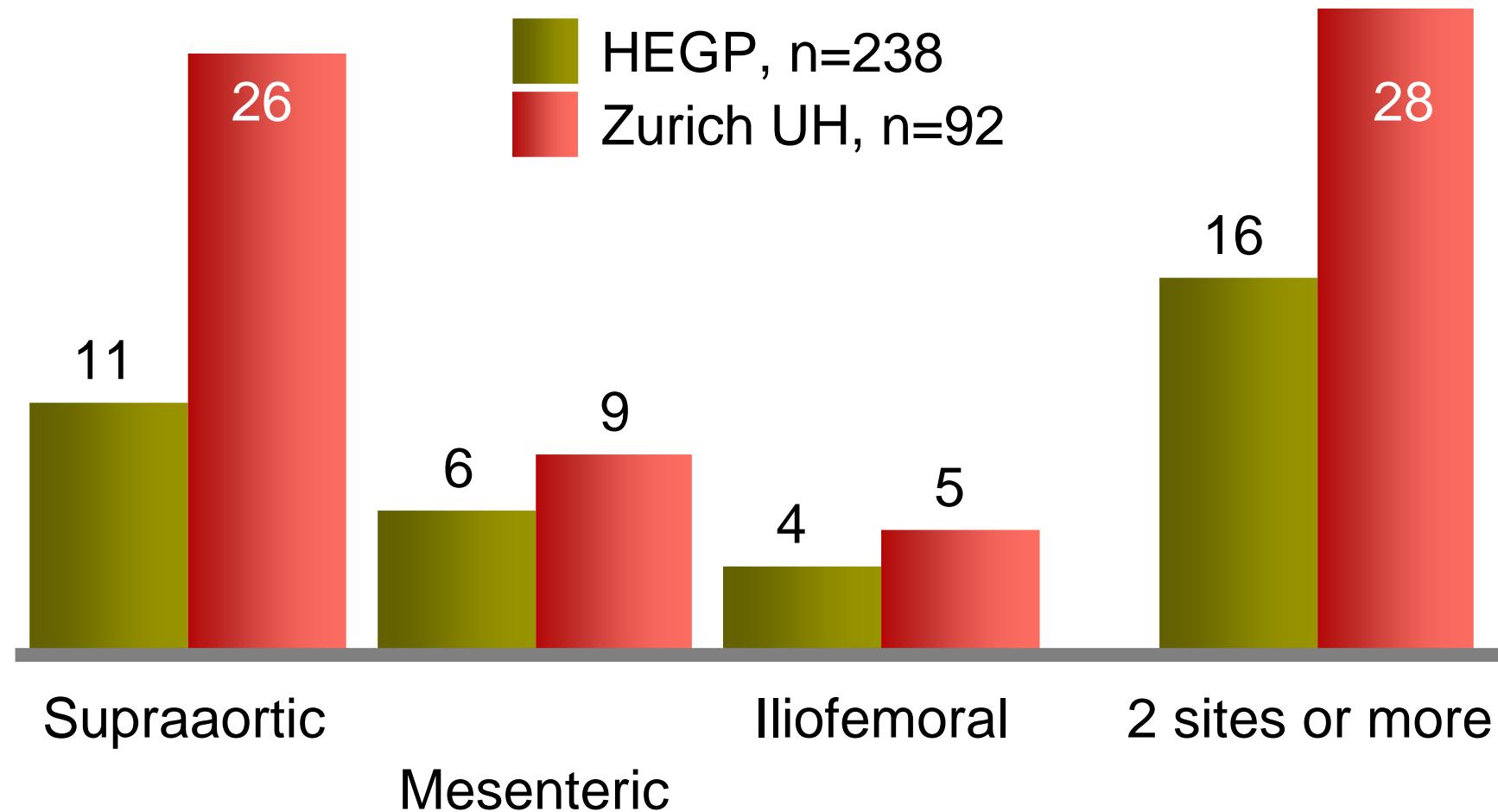


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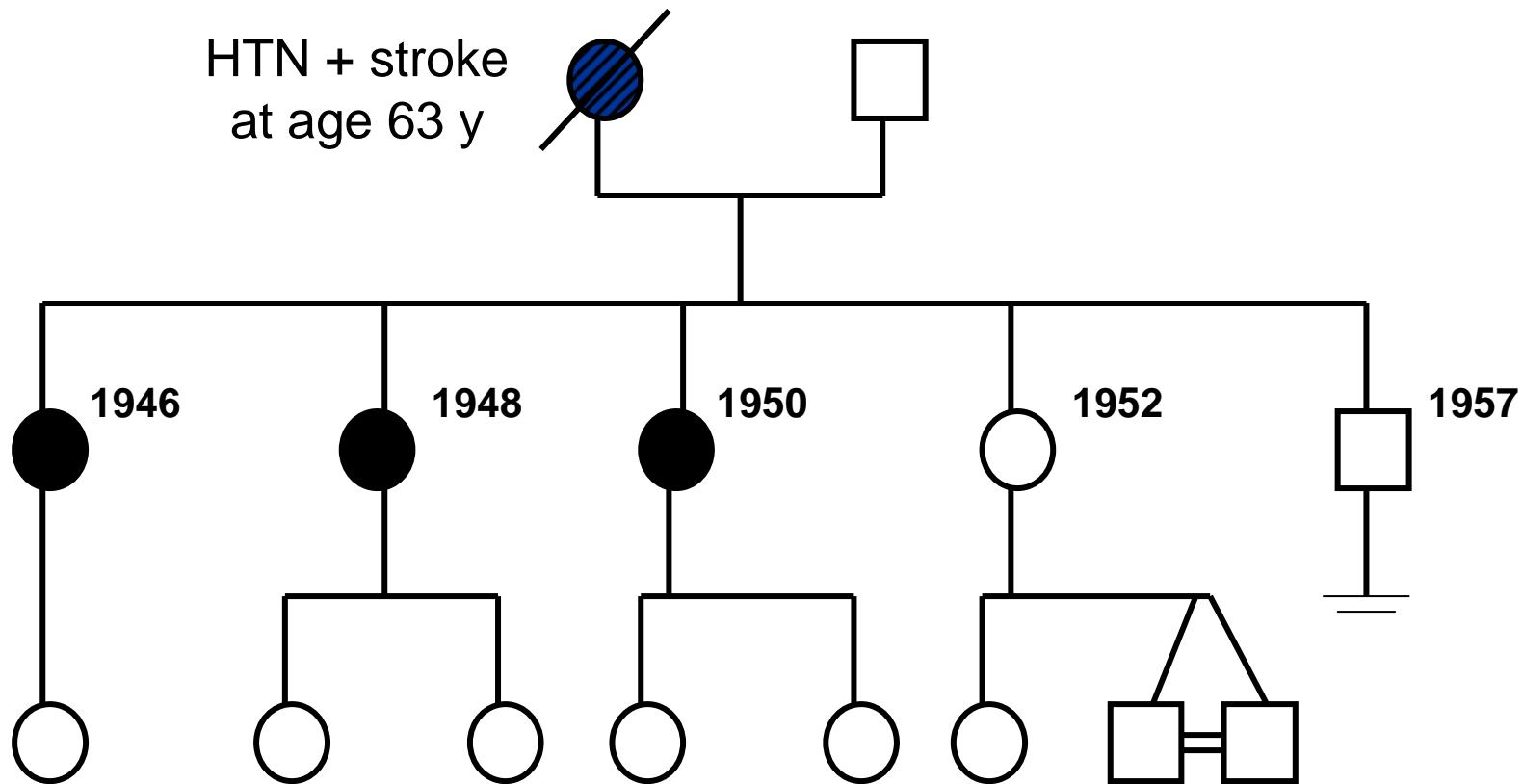
Extrarenal lesions (%) in renal artery FDM



Luscher TF et al, Nephron 1986;44 (suppl 1):109

One patient at HEGP and 2 at Zurich UH had coronary artery FMD

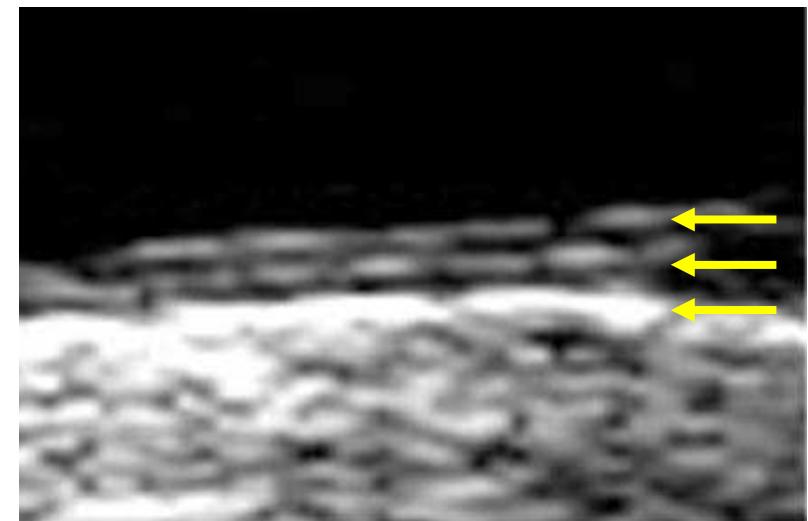
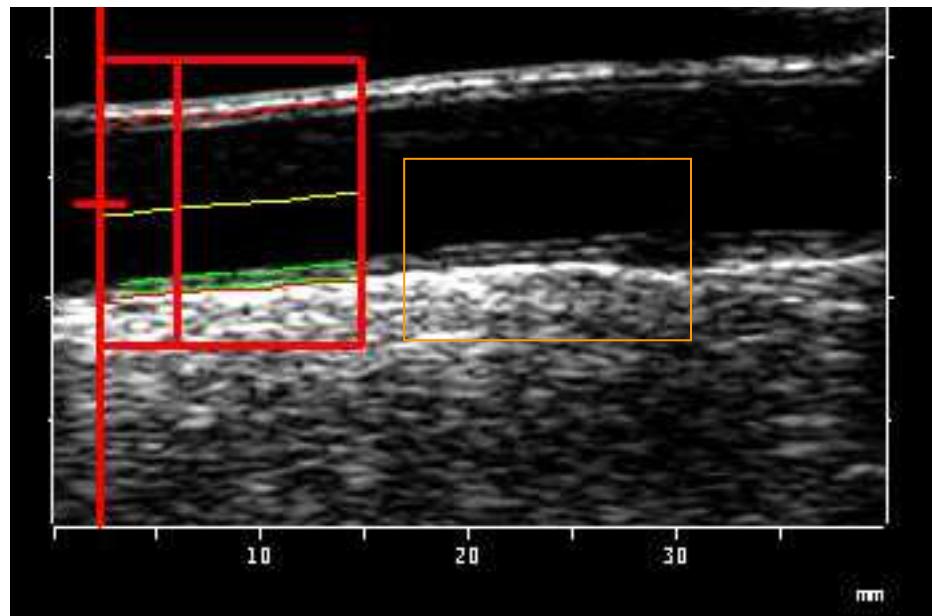
FMD may be familial* (OMIM #135580)



* Defined as the presence of angiographically documented FMD in at least 2 first-degree relatives

High-resolution echo-tracking in FMD

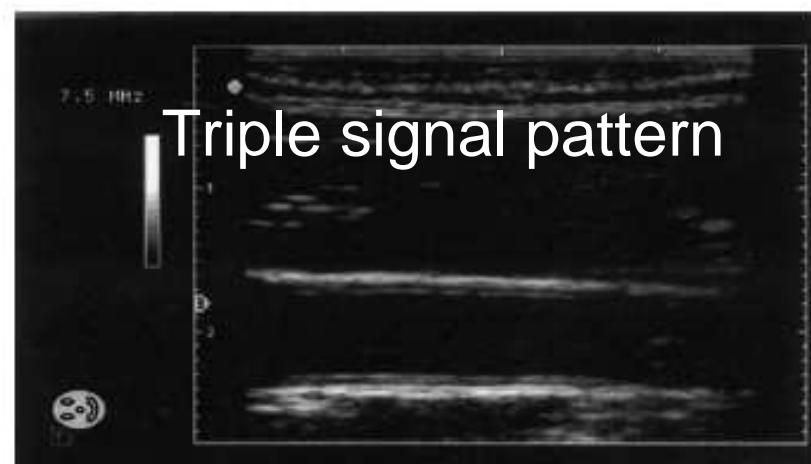
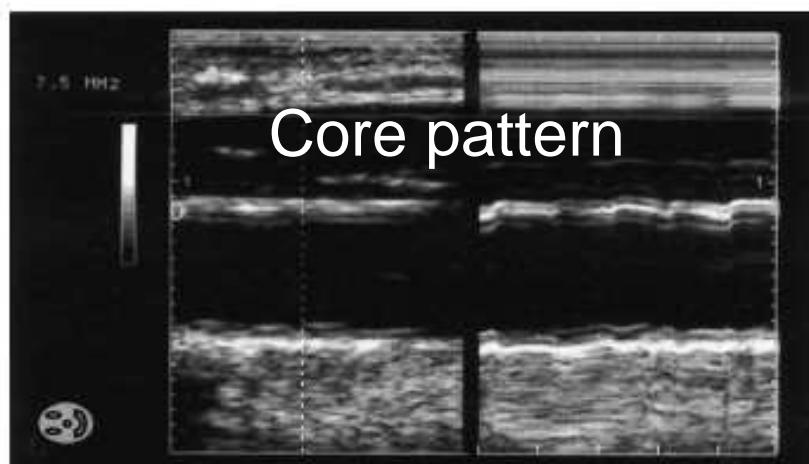
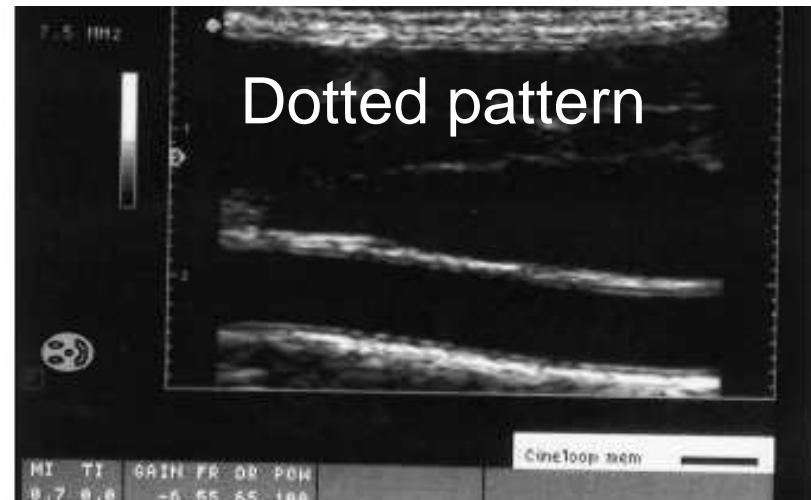
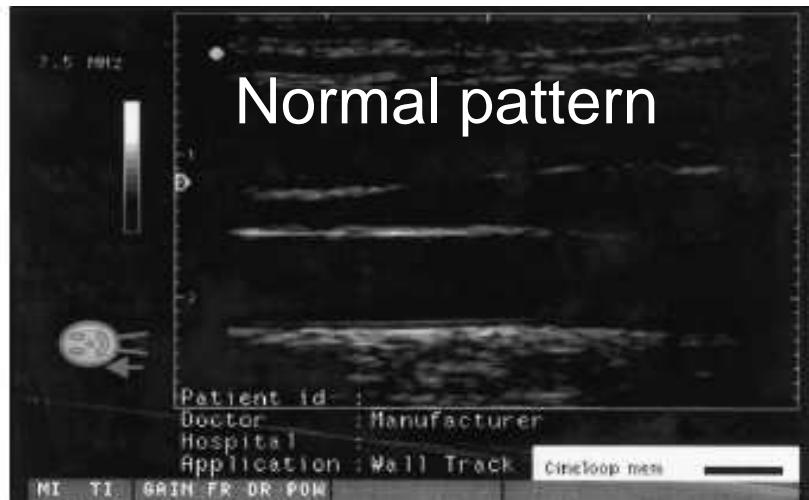
Patients with FMD had thicker common carotid (+12%) and radial arteries (+10%) than controls, and frequent abnormal echographic patterns of the carotid artery



x8 magnification
note the triple aspect of the distal wall

Boutouyrie P et al, J Hypertens 2003;21:2287

FMD is a systemic disease



Boutouyrie P et al, J Hypertens 2003;21:2287

Familial resemblance of arterial score

	sporadic cases, 125	familial cases, 13	relatives	controls
			47	47
Females, %	89	92	45	45
Bilateral FMD, %	58	77	-	-
Multifocal, %	74	100	-	-
BP, mmHg	177/105	186/112	129/76	127/74
Phenotypic score	4.0	4.8	4.2	2.5

Segregation analysis suggested an
autosomal dominant transmission of the trait

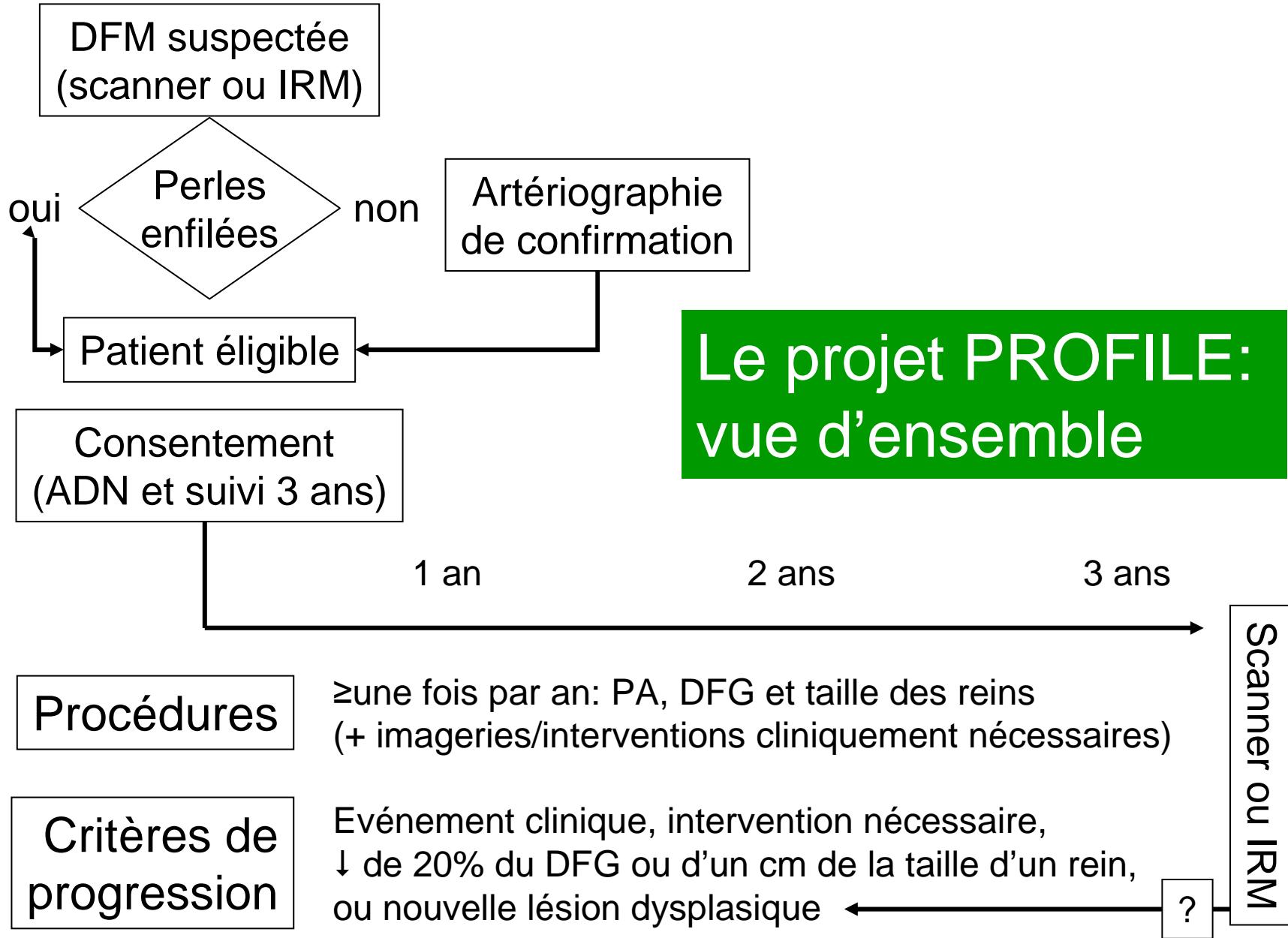
Perdu J et al, J Human Hypertens 2007;21:393

Fibromuscular dyplasia: conclusions

- FMD is a group of diseases which differ by patient's age, angiographic phenotype and stenosis severity
- The most prevalent type is associated the string of beads appearance (medial, multifocal FMD)
- The major aims of current research are:
 - to improve the quantification of renal artery FMD lesions
 - to assess more accurately the frequency of multi-site FMD and the risk of disease progression
 - to unravel the pathophysiological mechanisms of FMD and to seek gene(s) that predispose to the condition

Le projet PROFILE, objectifs

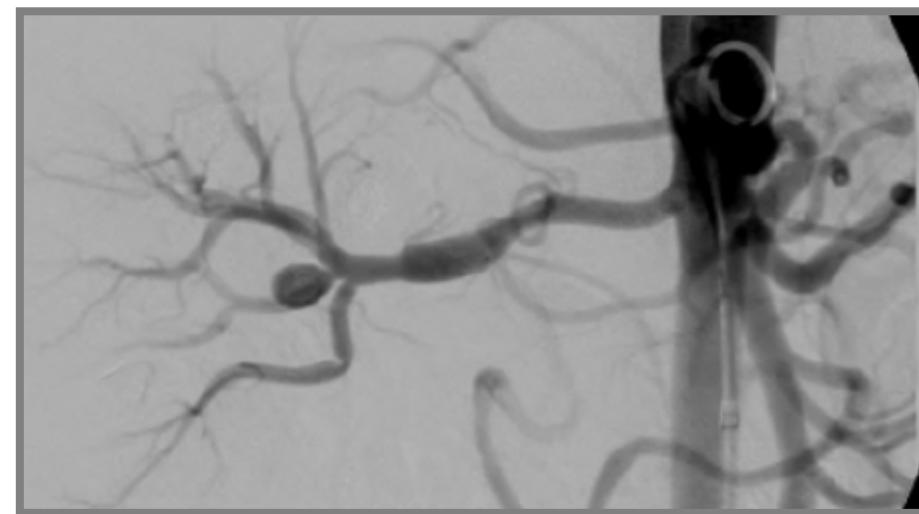
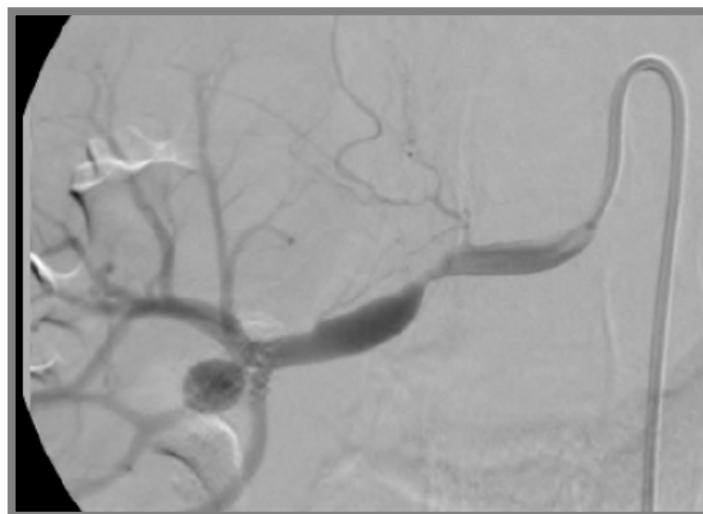
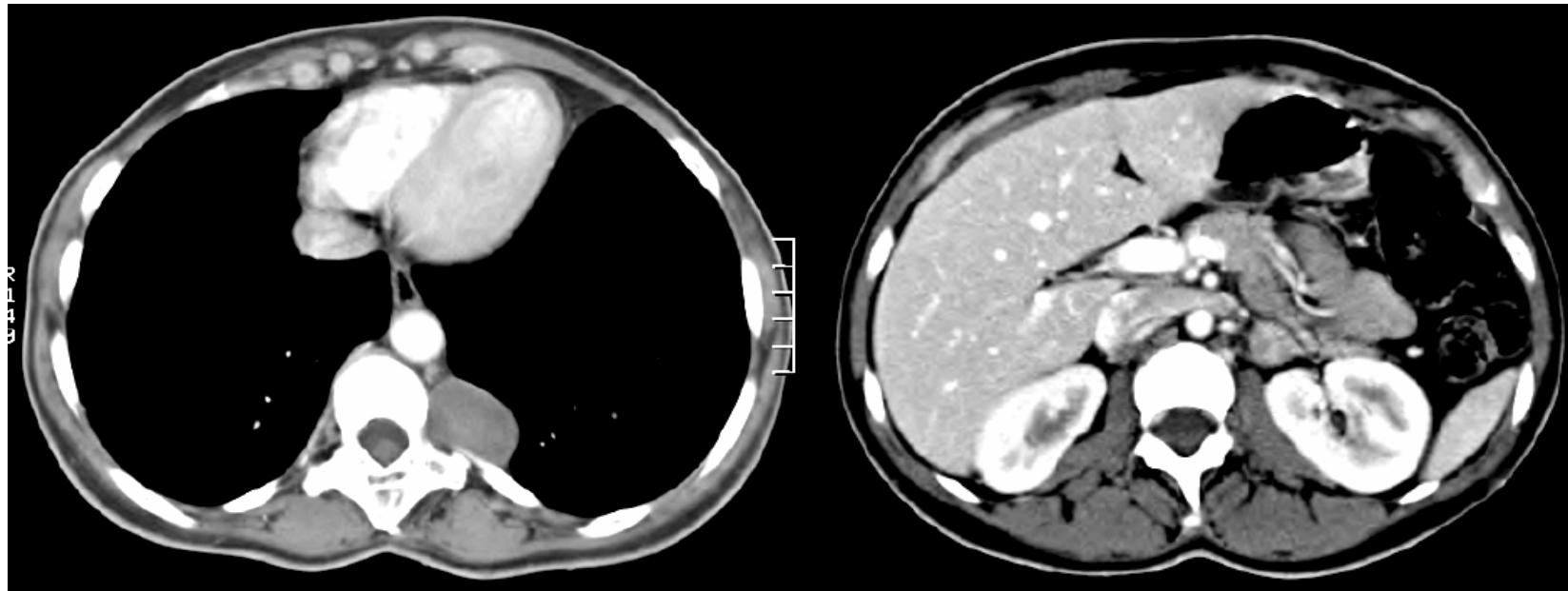
- Objectif principal: documenter l'incidence de la progression à 3 ans et ses facteurs de risque
- Objectif secondaire: documenter la fréquence des atteintes multiples et leurs facteurs de risque
- Moyens: créer une database (clinique, biologique et radiologique) et une biobanque pour une recherche coopérative sur les DFM



Causes rares

- Aux frontières de la dysplasie:
 - Neurofibromatose de type I
 - Ehlers-Danlos vasculaire, syndrome d'Alagille
- Sténose radique (athérosclérose expérimentale)
- Takayasu et autres maladies inflammatoires
- Hématome disséquant et infarctus rénaux

Dysplasie artérielle rénale dans la NF1



Syndrome d'Ehlers-Danlos vasculaire



Artérite de Takayasu: critères de l'ACR

1. Début avant 40 ans
2. Claudication des extrémités, notamment des MS
3. Armortissement d'au moins un pouls brachial
4. Anisotension ≥ 10 mm Hg
5. Souffle sur \geq une sous-clavière ou sur l'aorte abdominale
6. A l'artériographie, sténose ou occlusion de l'aorte, de ses branches ou des artères de grand calibre des membres, en l'absence d'athérosclérose et de dysplasie; les atteintes sont habituellement focales ou segmentaires

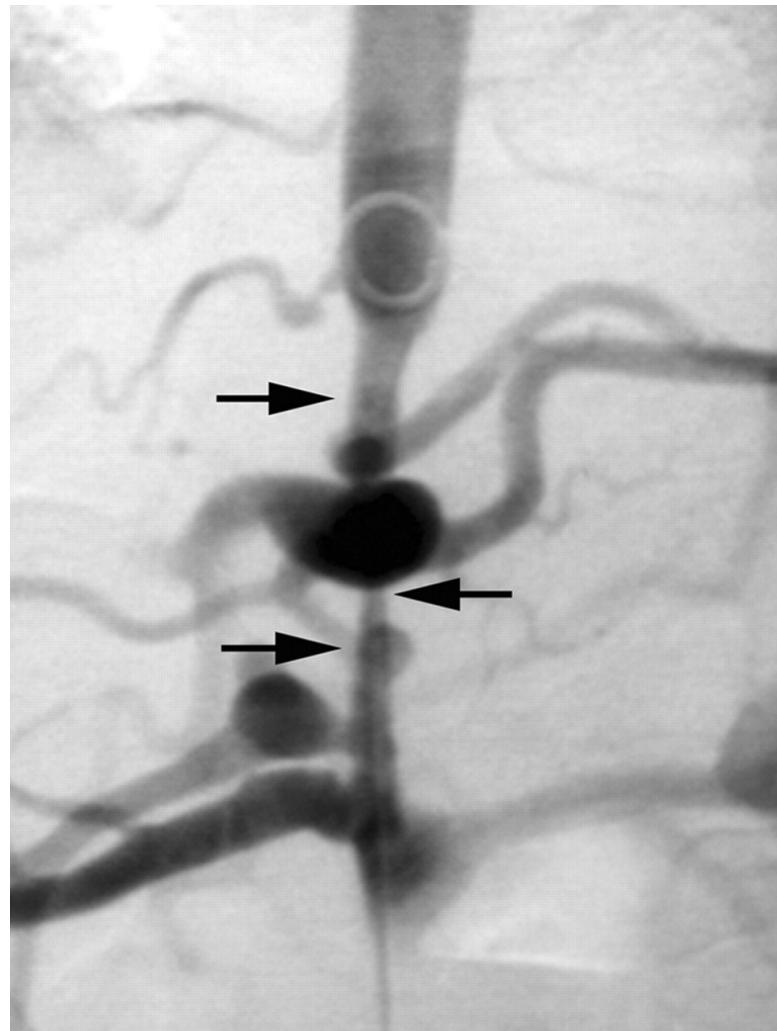
Le diagnostic requiert au moins 3 des 6 critères

32-year-old woman with Takayasu's arteritis



Gotway MB. et al. Am J Roentgenol 2005;184:1945

Aorto-artérite de Takayasu



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Cardiol

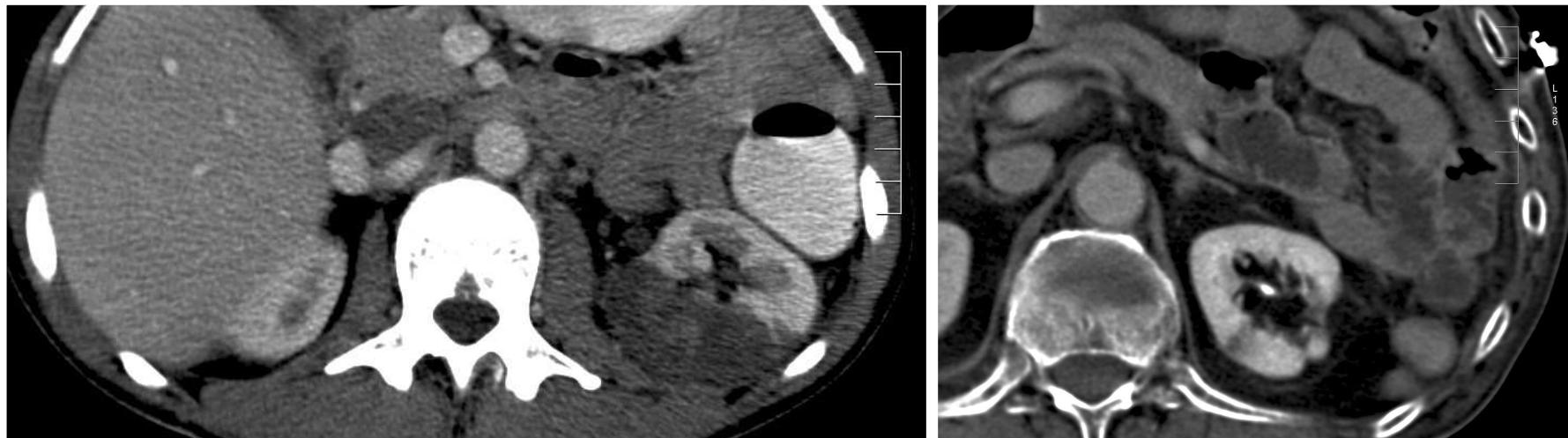
Gotway MB. et al. Am J Roentgenol 2005;184:1945

FACULTÉ
DE MÉDECINE
PARIS DESCARTES


Unité d'hypertension, HEGP

Infarctus rénal

- Plus souvent lié à une lésion artérielle rénale qu'à une embolie systémique
- Il se manifeste typiquement par une lombalgie avec ou sans hématurie, avec une écho normale
- Il peut causer une HTA sévère, parfois réversible



Paris B et al. J Hypertens 2006 ;24:1649