

# Spontaneous Coronary Dissection and Its Long-Term Prognostic Implications in a Cohort of 19 Cases

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Spontaneous coronary artery dissection is a rare cause of acute coronary syndrome. It mainly affects women with no significant cardiovascular risk factors and its presentation varies from unstable angina to sudden death. Knowledge of the condition is based only on individual case reports and the lack of large case series means that its treatment and prognostic implications have not been fully established. We present data on 19 instances of spontaneous coronary artery dissection in 18 patients who were treated at our center between May 1998 and January 2009. The median follow-up period was 3.8 years (interquartile range: 1.3-4.6 years). Once the acute phase had passed, the prognosis was favorable and there were no implications for functioning. One patient presented with a relapse in another coronary artery and another patient gave birth without complications 3 years after the dissection.

**Key words:** *Coronary artery dissection. Coronary angiography. Myocardial infarction. Prognosis.*

## Dissección coronaria espontánea y sus implicaciones pronósticas a largo plazo en una cohorte de 19 casos

La disección coronaria espontánea es una causa infrecuente de síndrome coronario agudo que afecta predominantemente a mujeres sin apenas factores de riesgo cardiovascular y cuya presentación varía desde la angina inestable hasta la muerte súbita. Los conocimientos de esta entidad se reducen a casos clínicos aislados, y la ausencia de grandes series hace que su tratamiento y sus implicaciones pronósticas no estén plenamente establecidos. Presentamos los datos de 19 casos en 18 pacientes atendidos en nuestro centro desde mayo de 1998 hasta enero de 2009, con una mediana [intervalo intercuartílico] de 3,8 [1,3-4,6] años de seguimiento. Una vez superada la fase aguda, el pronóstico fue favorable sin implicaciones funcionales. Una paciente presentó una recidiva en una coronaria diferente y otra paciente dio a luz sin complicaciones a los 3 años de la disección.

**Palabras clave:** *Disección coronaria. Angiografía coronaria. Infarto de miocardio. Pronóstico.*

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## INTRODUCTION

Spontaneous coronary artery dissection (SCAD) is a rare cause of acute coronary syndrome (ACS).<sup>1</sup> It mainly affects women who have few cardiovascular risk factors and its diagnosis has increased as the use of coronary angiography has expanded. SCAD is the separation of 2 of the 3 arterial wall layers,<sup>1</sup> creating

a false lumen<sup>2</sup> whose progression can impair distal coronary blood flow and cause ischaemia, with various presentations ranging from elevated cardiac markers and ST without pain to sudden death.<sup>1</sup> Our understanding of this syndrome is limited to a few isolated case reports and some small series and therefore its treatment and prognosis are not well established.<sup>3</sup>

## METHOD

We analysed 19 cases of primary SCAD diagnosed in a tertiary hospital from May 1998 until January 2009. In this period, our centre performed 9502 angiograms on patients with ACS, which has increased considerably in the last 5 years. This is a descriptive study with follow-up.

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**TABLE 1. General Epidemiological Characteristics**

Total cases in 18 patients	19
Age, mean (SD), y	47.7 (10.4)
Female, n (%)	15 (79)
Arterial hypertension n (%)	7 (37)
Dyslipaemia n (%)	4 (21)
Diabetes mellitus n (%)	2 (10)
Current smokers, n (%)	1 (5)
Former smokers, n (%)	6 (31)
Body mass index, mean (SD)	26.6 (5.4)
Affected artery	
ADA, n (%)	12 (63)
ACx, n (%)	5 (26)
RCA n (%)	2 (11)
Maximum troponin T, mean (SD), ng/mL	1.46 (0.8)
Previous treatment	
Antihypertensive, n (%)	7 (37)
Statins n (%)	4 (21)
Hormones, n (%)	7 (37)

ACx indicates anomalous circumflex artery; ADA, anterior descending artery; RCA, right coronary artery; SD, standard deviation.

The 2 cases in the same patient have been analyzed as one sole case.

## RESULTS

The epidemiological characteristics of the series are shown in Table 1, while the individual breakdown of clinical presentation and the subsequent course of the disease are shown in Table 2. All cases in which ST elevation was documented underwent fibrinolysis or were entered into a primary angioplasty programme

at our center, despite the fact that stent implantation was rejected (considered a conservative posture in the Table).

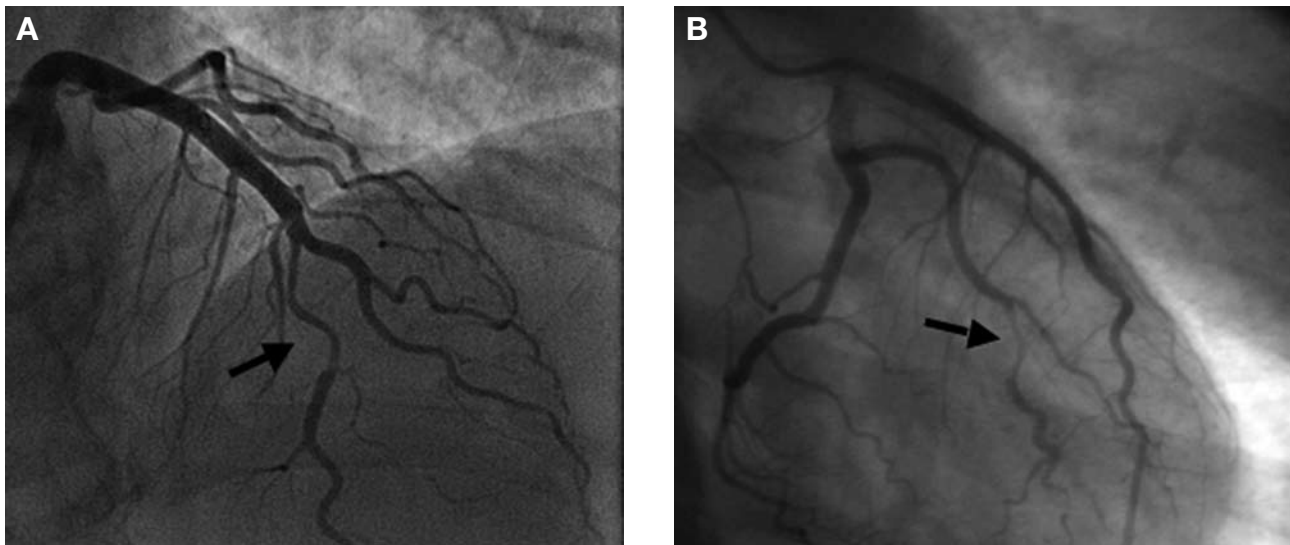
One of the cases appeared 2 days after uncomplicated vaginal delivery and 3 cases occurred in athletes after exhausting physical exertion (24-48 hours after exertion, with 2 of these cases after having run a marathon). Intense stress during the previous 24 hours was noted in 5 cases, and 7 of the cases occurred in patients taking estrogen (3 for oral contraception and 4 for perimenopausal hormone replacement therapy). The presence of autoantibodies was ruled out (in all cases: ANCA, ANA, and anti-DNA; in cases occurring in the last 5 years also anticardiolipin). No arteriosclerotic lesions were detected at other levels in the coronary angiographies (Figure 1). On 3 occasions, the diagnosis was confirmed using intravascular ultrasound.

The first case in the series deserves special attention: a 47-year-old woman whose sister had spontaneous dissection of the internal carotid artery presented 2 SCAD (case 1 and 2 of the series). The first episode occurred 24 hours after running a marathon, and she was on oral contraceptives at the time. The patient presented with midthoracic tightness accompanied by intense vegetative symptoms. The emergency team determined anterior ST elevation and she underwent fibrinolysis. The following day, a catheterization was performed that revealed an anterior descending artery dissection in the middle segment and, due to her clinical stability,

**TABLE 2. Clinical Characteristics and Patient Outcomes**

Case	Sex	Age	ACS	Symptoms	Factor	Artery	Intervention	EF	Follow-up
1	F	47.8	ACSWSTE	A + VS	Sports + OC	ADA m	Fibrinolysis	60	2. SCAD
2 (1B)	F	51.4	ACSWSTE	A + VS	Stress	RCA p	Fibrinolysis + stent	58	Good
3	F	35.7	ACSWSTE	A + VS	OC	ACx d	Conservative	60	Good
4	F	37.9	ACSWSTE	A + Ar		ADA p	primary PTCA	68	Good
5	F	50.7	ACSWSTE	A	HRT	ADA m	Conservative	55	Good
6	F	31.2	ACSWSTE	A	Pregnancy	ADA p	primary PTCA	67	Good
7	F	55.9	ACSWSTE	A + VS		ADA d	Conservative	68	Good
8	F	34.4	ACSWOSTE	A + Ar	OC + tabacco	ACx d	Conservative	65	Preg
9	F	45.7	ACSWOSTE	A		ACx p	Stent	65	Good
10	F	53.1	ACSWSTE	A		ACx p	Conservative	60	Good
11	M	42.2	ACSWSTE	A	Sports	ADA l	Conservative	60	Good
12	M	42	ACSWOSTE	A + VS	Stress	ADA p	Stent	65	Good
13	M	49.6	ACSWOSTE	A	Stress	ADA m	Conservative	65	Good
14	F	50.7	ACSWOSTE	A	HRT	ADA p	Stent	65	Good
15	M	45.2	ACSWOSTE	A	Sports	ADA m	Conservative	70	Good
16	F	40.2	ACSWSTE	A	HRT	ADA p	Conservative	68	Good
17	F	52.2	ACSWSTE	A	Stress	ACx p	Conservative	65	Good
18	F	57.1	ACSWSTE	A	HRT	RCA m	Conservative	68	Good
19	F	74.1	ACSWSTE	A + VS	Stress	ADA m	Fibrinol+cons	50	Good

A indicates angina; ACS, acute coronary syndrome; ACSWSTE, ACS with ST elevation; ACSWOSTE, ACS without ST elevation; ACx, anomalous circumflex artery; ADA, anterior descending artery; Age, years at admission; Ar, arrhythmias; d, distal; EF, ejection fraction at discharge; F, female; Factor, trigger event; Fibrinol+cons, patient underwent fibrinolysis at another center and was then moved to our center for catheterization, without stent implantation; HRT, hormone replacement therapy; m, median; M, male; OC, oral contraceptives; p, proximal; RCA, right coronary artery; SCAD, spontaneous coronary artery dissection; VS, vegetative symptoms.



**Figure 1.** Diagnostic coronary angiographies. A: spontaneous coronary dissection of the anterior descending artery in postpartum (case 6). B: typical narrowing of coronary dissection in a distal segment of the first marginal branch of the circumflex artery (case 8).

conservative treatment was chosen. Four years later, the patient still practiced sports regularly and had stopped taking contraceptives. Being on duty and after having treated a youth suffering from cardiorespiratory arrest, the emergency service attended to her. She presented similar symptoms with a new ST elevation, this time lower, and with an incomplete response to the initial fibrinolysis. In the subsequent catheterization, a proximal right SCAD was observed and a regular stent was implanted. The previously affected descending anterior artery did not have angiographic disorders this time around.

None of the patients underwent heart surgery and they were all discharged with preserved ejection fraction and given aspirin, beta-blockers and angiotensin-converting enzyme inhibitors or angiotensin II receptor antagonists as part of their treatment.

During outpatient follow-up, there were no cardiovascular events, regardless of treatment, except in the previously mentioned first case and no subsequent collagenopathies or neoplasias that might have been related to these events were diagnosed. The cardiologist who monitored each patient decided the long-term treatment unilaterally. Long-term use of aspirin was prescribed for cases with stent implants and for one case that later developed diabetes; beta-blockers were prescribed for 14 patients and low-dose statins were prescribed for 9 patients.

The patient in case 8 became pregnant and gave birth to a healthy baby who was delivered vaginally without complications two years after the dissection. The patient discontinued oral contraceptives and smoking after the heart attack and now engages in sports on a regular basis and is so far symptom-free.

## DISCUSSION

SCAD is a rare disorder involving the dissection of the coronary wall, usually between the intima and the media (in cases involving atherosclerosis) or between the media and the adventitia (mostly in cases associated with childbirth). Ever since the first case was diagnosed in 1931 there have been over 300 published cases, many from autopsies performed after sudden death.<sup>2</sup> The data are consistent with those of our series: more than 70% of those affected are women, 30% of cases are related to pregnancy, the anterior descending artery is affected in more than half of the cases and, once the acute phase is overcome, SCAD has a good long-term prognosis.<sup>3</sup>

The pathophysiology is not fully understood and has been associated with the inflammatory rupture of atherosclerotic plaques. In cases related to childbirth and/or postpartum, pericardial eosinophilic infiltrates have been reported during autopsies.<sup>2,4</sup> This could be related to the effect of oestrogen and progesterone stimulating the eosinophilic secretion of granules with lytic substances.<sup>4</sup> This may also occur as a consequence of female hormonal treatments, which have already been associated with this problem.<sup>1,2</sup>

Other factors related to SCAD include hypertension,<sup>5</sup> cocaine use,<sup>2</sup> coronary spasms due to increased shear forces in the vessel,<sup>2</sup> vasculitis,<sup>6</sup> syndromes that present with abnormalities in the vascular walls (e.g., Marfan<sup>7</sup>), treatments such as cyclosporine<sup>2</sup> and coronary compressive forces secondary to intramyocardial pathways and exhausting physical exercise.<sup>8</sup> It is worth mentioning that there is a possible hereditary factor at play that could explain the reported familial groupings,<sup>9</sup> such as with the first patient in our series.

Widespread use of coronary angiography in ACS has increased the number of diagnosed cases. An intimal flap is often observed, appearing as a radiolucent line that separates the double lumen along with indirect signs of dissection, such as the retention of contrast in the false lumen related to stenosis (Figure, A).<sup>3</sup> Less often, we find arterial narrowing with no evidence of a flap, when there is no contrast flow in the false lumen (Figure, B). If in doubt, intravascular ultrasound<sup>10</sup> and, even in the initial stages of implantation, optical coherence tomography<sup>11</sup> can assist with a definitive diagnosis, especially when it is necessary to verify that the guidewire is in the true lumen before proceeding with the intervention. Coronary computed tomography<sup>12</sup> can also help clarify the diagnosis non-invasively, especially when complications are suspected.

Treatment regimens depend on the location of the dissection, its accessibility and extent, the severity of the ischaemia and patient stability.<sup>2</sup> The decision should be made on a case-by-case basis. For stable patients, those with only a small affected vessel or those whose affected vessel is in a location that precludes intervention, conservative medical therapy should be chosen. This happened in more than half of the cases in our series and is supported by the spontaneous resolution of the dissection during follow-up, as occurred in our first case. In patients with ischemia, especially those with severe affectation near a vessel, percutaneous intervention with direct stent implantation is recommended. For cases of multivessel disease and particularly when the left main trunk is affected, cardiac surgery may be the therapy of choice. There are reported cases where trunk dissection has been conducted successfully and conservatively.<sup>13</sup>

Fibrinolysis is not recommended due to risk of increased bleeding in the heart wall and advancing the dissection.<sup>1,3</sup> Fortunately, it is applied less and less often as primary angioplasty programs are available 24 hours a day.

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