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Information sheet

European Society of Cardiology European Position Paper on SCAD

Below is Beat SCAD's summary of the European Society of Cardiology, acute cardiovascular care association, SCAD study group Position Paper on Spontaneous Coronary Artery Dissection (SCAD).

The Position Paper (Published February 2018) explains current knowledge of SCAD for healthcare professionals and provides a consensus on management, areas of controversy and uncertainty https://bit.ly/2DH9Lzm.

The information below is a summary of the main points in the Position Paper. We have tried to explain as much of the Paper as possible in plain English for SCAD patients and their families.

We highly recommend GPs, cardiologists, cardiac rehab staff and other health professionals to read the full paper and/or the summary in Openheart (https://bit.ly/2zd1500).

Please note, this information sheet is not intended as medical advice and SCAD patients should always discuss their treatment with a medical professional.

SCAD used to be considered rare and associated mainly with pregnancy and post-partum, however higher sensitivity troponin tests and better imaging techniques, as well as a greater awareness of SCAD, has led to increased diagnosis, with the result that SCAD is now seen as a significant cause of Acute Coronary Syndrome (ACS) in young to middle-aged women and the minority of cases are pregnancy related.

There are key differences in management and outcomes compared to atherosclerotic heart disease (where plaque builds up in the arteries).

In SCAD a false lumen (passageway) develops in an artery wall, into which blood flows and compresses the true lumen, reducing blood flow. There are two theories about the cause of this: 'inside-out' and 'outside-in'.

'Inside-out': the development of a 'tear', allowing blood to accumulate in the artery.
'Outside-in': the development of a 'bruise' that haemorrhages (leaks) into the artery wall.

Incidence & demographics

It is unknown how frequently SCAD occurs. Current figures from various studies across the world show a SCAD diagnosis in 0.07-0.2% of all angiograms and 2-4% of angiograms performed for ACS. Pregnant/post-partum cases account for around 10% of cases of SCAD overall, however 21-27% of heart attacks in pregnancy and 50% of post-partum coronary events are reportedly due to SCAD. SCAD has been reported in patients aged 18-84, with the average between 44 and 53. No ethnic variations have been reported but there is a strong female predominance.

Risk factors & associations

The paper says: "It is likely that a combination of predisposing factors increase susceptibility" so a relatively minor event can trigger a SCAD.

Approximately 90% of SCAD patients are women, suggesting a link with female sex hormones.

Other reported associations include pregnancy and Fibromuscular Dysplasia (FMD).

Some evidence from Canada indicates that male SCAD patients are different from female cases – slightly younger, with higher rates of isometric exercise (exercises that involve holding a position rather than moving, eg plank) and lower emotional stress levels prior to SCAD.

P-SCAD (pregnancy related) SCAD happens in around 10% of cases. Most SCADs happen during the third trimester. Post-partum SCADs have been reported up to 2 years after the birth, but most are reported during the first six weeks.

A Canadian study suggests the P-SCAD may be more severe than other types of SCAD.

FMD, a "non-atherosclerotic, non-inflammatory disease of arterial walls", has two sub-types: multifocal (artery looks like a 'string of beads' and (uni)focal (artery narrows – often). Studies have reported FMD in SCAD patients in 11-86% of patients. The range narrows to 41-86% after excluding three studies where less than 50% of patients were screened. Prevalence of FMD may differ depending on the proportion of patients screened and the screening method used. FMD may be more frequent in SCAD patients with more tortuous (curvy/winding) coronary arteries.

The most commonly affected arteries are renal (kidney), cervico-cephalic (neck and head) and iliac (pelvis).

Other than FMD lesions (damaged areas), other vascular abnormalities such as aneurysms, dissections, irregularities, undulations and/or tortuosity in vessels outside the heart have been reported in SCAD patients.

The paper warns that although it's tempting to see SCAD as a complication of FMD, a degree of caution is required as the current data about SCAD and FMD mostly comes from two centres in Canada and the USA, typical FMD coronary lesions are rare, the prevalence of coronary artery dissection is very low in the US FMD registry (<3%) and some SCAD cases are idiopathic (the cause is unknown/not associated with FMD) or associated with other vascular diseases.

Atherosclerotic risk factors

SCAD patients typically have few or no traditional risk factors for atherosclerotic (plaque-induced) heart disease. Some patients do, however, have some risk factors, including high blood pressure, smoking and high cholesterol, although there is no evidence these contribute directly to SCAD.

Mechanical stressors and exercise

Some mechanical triggers have been linked to SCAD, including extreme Valsalva-type manoeuvres (holding your breath while lifting) and provocation of coronary spasm (sudden tightening of the muscles within the arteries of your heart). Isometric or extreme exercise have been reported in 11.9% of cases, most commonly in men.

Emotional stressors
Recent emotional stress, such as bereavement or personal crisis, have been reported in a higher than expected proportion of SCAD cases, particularly in women.

Genetics
A few sibling-sibling and mother-daughter pairs of SCAD patients have been reported, but SCAD does not appear to be a strongly inherited condition, with only 1.2% of 412 patients in one study reporting a family history.

Clinical presentation
There is a lot of evidence that SCAD is under-diagnosed due to patients not acting on symptoms, sudden cardiac death where post-mortems don't find SCAD, as well as missed or delayed diagnosis due to patients having low risk factors for atherosclerotic heart disease.

Chest pain is the most frequent symptom (60-90% of patients). In the Canadian series chest pain was associated with pain in the arm (49.5%), neck (22.1%), nausea and vomiting (23.4%), excessive sweating (20.9%), laboured breathing (19.3%) and back pain (12.2.%).

Diagnosis
Currently there is no blood test that can identify SCAD.

Other conditions that often get considered before a SCAD diagnosis is made include:
Atherosclerosis (plaque)
Coronary artery spasm
Takotsubo cardiomyopathy
Coronary thromboembolism
Myocardial infarction with non-obstructed coronary arteries (MINOCA)

Coronary angiography is the main way to diagnose SCAD, although many interventional cardiologists are not used to spotting the signs of SCAD on angiograms. Intracoronary imaging (eg CT scan) is done where there is uncertainty about diagnosis.

There are four types of SCAD:
Type 1 SCAD: a 'flap' and 'linear double lumen' – 29-48% of cases.
Type 2 SCAD: a long diffuse (occurs over a wide area as opposed to a specific location) and smooth narrowing mainly in the mid-to-distal segments – 52-67% of cases.
- **Type 2a**: where an artery further away from the SCAD has a normal diameter.
- **Type 2b**: where the narrowing extends to the end of the artery.

Type 3: a narrowing that is indistinguishable from an atherosclerotic narrowing (intracoronary imaging required to confirm) – 0-3.9% of cases.
Type 4: complete blockage usually of a distal (further away from the top of the heart) artery.

There may be: increased tortuosity of the arteries or coronary FMD, SCAD may occur in more distal vessels compared to atherosclerotic heart disease, the LAD (left anterior descending) is reported to be the main artery affected, there is no or very little atherosclerosis.

Dissections in more than one artery simultaneously reportedly occurs in 5-13% of cases.

One study has reported that there is an increased risk of a further dissection during angiography (2% risk compared to 0.2% in non-SCAD cases) and PCI (percutaneous coronary intervention – 'stenting') (14.3%). Avoidance of "aggressive or deeply engaging guiding catheter designs is advised".

Most SCADs can be diagnosed with angiography and if possible a conservative approach to treatment is recommended.

Careful intracoronary imaging appears safe. Intravascular ultrasound (IVUS) and Optical coherence tomography (OCT) are valuable in diagnosing SCAD.

Given the increased risk of dissections caused by coronary angiography and angioplasty, repeat imaging should only be considered when clinically necessary.
Computer tomography coronary angiography (CTCA) is non-invasive, so does not risk dissections caused by invasive angiography (see previous page), however is limited due to resolution and false negatives have been reported. It is recommended, therefore that the primary diagnostic tool should be coronary angiography, with CTCA used as a follow-up.

Acute management

Evidence suggests the majority of SCADs will heal over time if managed conservatively (ie with medicine only). Where conservative management fails, the majority of cases occur during early follow-up, so monitoring in hospital for around five days is advised.

Repeat angios are challenging due to underlying "disrupted and friable (fragile) coronary vessel wall" and where repeat angios are not needed, conservative management is favoured.

Studies show an increased risk of complications with PCI (stenting). Risks include increased risk of a secondary dissection, guidewire entering the false lumen, the false lumen increasing in length when stents are inserted, persistent distal dissection, blockage of a side branch due to the haematoma (bruise) getting larger. Conservative management is therefore recommended unless the patient is unstable and requires intervention. Where stents are used, second-generation drug eluting stents are advised.

Coronary artery bypass outcomes are reported to be good, but one study reported high rates of graft failure, possibly due to healing of the dissected artery, which starts working again so the graft is no longer required.

Medical management

There are no randomised controlled trials to compare different conservative treatments so current practice is based on guidelines for non-SCAD ACS treatment.

Thrombolysis (treatment to dissolve blood clots) should not be used for acute management of SCAD.

Antiplatelet therapy (which reduces risk of blood clots) is controversial for various reasons, one of which is the concern about using medication that extends bleeding time for a condition that may be an intramural bleed (a bleed between the walls of the artery). Also, giving patients of menstrual age antiplatelet medication could cause menorrhagia (abnormally heavy bleeding during periods). Patients who have stents should be on dual antiplatelet therapy (typically aspirin and clopidogrel) for 12 months and "prolonged or lifelong monotherapy (usually with aspirin)". Conservatively managed patients are advised to have dual antiplatelet therapy in the 'acute' phase of recovery. The optimal duration of antiplatelet therapy is unknown and some clinicians question the approach that calls for lifelong aspirin.

Anticoagulant therapy (blood thinners) – there are concerns about the same issues as antiplatelet therapies (see above) and anticoagulant therapy should probably be limited to the acute period unless there is a clinical need (eg a blood clot).

ACE inhibitors (blood pressure medication), angiotensin receptor blocker (ARB) (to dilate blood vessels and reduce blood pressure), mineralocorticoid receptor antagonists (MRA) (a diuretic), betablockers (to lower heart rate and therefore blood pressure) and vasodilator (dilates blood vessels to lower blood pressure) therapies – current guidelines for ACE, ARB and betablockers should be followed for SCAD patients with significant impairment of left ventricular systolic function*, adding MRA (diuretic) as necessary. Low blood pressure in some SCAD patients can limit increasing the dosage. However, prescribing these for patients without significant left ventricular systolic function impairment is controversial. One study has linked high blood pressure with an increased risk of

recurrent SCAD and betablocker treatment with reduced risk of recurrence. If these results are validated it may provide evidence that SCAD recurrence risk may be reduced with medication. Nitrates or calcium channel blockers (vasodilatory therapies that dilate the blood vessels) can be used to treat recurrent chest pain.

Statins should be given to patients with high cholesterol.

Contraception and hormone replacement therapy – concerns are based on the assumption that there is an association between SCAD and female sex hormones, however it is still unclear what this association is. It would be reasonable to avoid hormonal contraception where possible. In patients with recurrent cyclical chest pain, low-dose local hormone delivery via IUD (typically the mirena coil) has been anecdotally reported to be useful.

Pregnancy post-SCAD

Some studies recommend SCAD patients should not become pregnant. There is limited data on the risk of pregnancy in SCAD patients but it should be considered high risk.**

Follow-up imaging

After SCAD, assessing the left ventricular systolic function is mandatory, using Echocardiography/ cardiac magnetic resonance imaging.

Imaging of extra-coronary vascular beds (blood vessels elsewhere in the body, ie outside the heart) is advised, given the association with FMD and other vascular conditions.

Recurrence

In patients surviving SCAD, long-term mortality is low.

A US study reported 17% recurrence rate across a period of 47 months, with a 10-year recurrence rate of 29.4%. A Canadian study reported 10.4% of 327 patients followed up for an average of 3.1 years. The Japanese study reported 7 recurrent SCADs after the first 30 days from 63 patients followed up for 34 months. The Swiss series reported 3 out of 63 recurrent SCADs followed up for an average of 4.5 years. The Italian study reported 4.7% recurrence over an average 22 months follow-up.

Recurrence often affects different arteries to the previous SCADs.

Reported recurrence rates may be overestimated due to the selection bias inherent to self-referral based studies.

Controlling hypertension (high blood pressure) may well reduce the likelihood of recurrence.

Post-SCAD chest pain

Recurrent chest pain is common after SCAD and sometimes happens cyclically, usually pre-menstrually.

Given the recurrence risk, patients should be assessed with ECG and troponin tests, but given the risk of dissection using invasive angiography, this should be used only when clinically necessary.

Anecdotally, symptoms may respond to vasodilator treatments that reduce vasospasms. Cyclical symptoms may respond to low dose contraception (eg progesterone hormonal coil).

Cardiac rehab and exercise

Several studies have shown that cardiac rehab exercise is safe and beneficial to SCAD patients and no study has found a link between exercise and recurrent SCAD. Extreme and isometric exercise are not advised, but given the benefits, both mental and physical, of exercise, “a return to full activity following SCAD, including non-extreme sport, is reasonable given the currently available data”.

Post-traumatic stress disorder (PTSD) and emotional/psychological consequences of SCAD

SCAD patients may be particularly at risk of PTSD and other adverse psychological issues as SCAD affects a younger, low-risk population where a life-threatening condition is unexpected. P-SCAD patients have the added challenge of a new baby. Cardiac rehab has been shown to be helpful and in some cases counselling, cognitive behaviour therapy (CBT), stress-reducing therapies or medication for anxiety or depression may be appropriate.

Beat SCAD Notes

* Measuring the left ventricular systolic function of your heart shows how well your heart is functioning. This is done by calculating the Ejection Fraction (EF). A normal left ventricular ejection fraction (LVEF) ranges from 55% to 70%. An LVEF of 65%, for example means that 65% of the total amount of blood in the left ventricle of the heart is pumped out with each heartbeat. Your EF can go up and down, based on your heart condition and how well your treatment works.
https://my.clevelandclinic.org/health/articles/16950-ejection-fraction

** For those wishing to discuss pregnancy options post-SCAD, we advise contacting a SCAD specialist for advice.