Chapter 4
Spontaneous Coronary Artery Dissection (SCAD): An Overview of the Condition, Diagnostic Work Up and Management

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Abstract There has been increased awareness of Spontaneous Coronary Artery Dissection (SCAD) as a type of myocardial infarction over the last decade. However, as its underlying pathology and mechanisms are still being understood, and the best management principles for SCAD still being realized, there are little robust guidelines for those not subspecializing in SCAD patient management to help guide management. This book chapter targets general medical practitioners who are faced with taking care of the small but increasing SCAD population in the community, in partnership with SCAD specialists. It provides an updated understanding of SCAD including the "inside out" versus "outside in" hypothesis of pathophysiology, and the management principles to be mindful of in SCAD in comparison to traditional acute coronary syndrome (ACS) management. Uniquely, the chapter provides colloquial answers to frequently asked questions by patients about SCAD, and an in-depth review of the benefits of cardiac rehabilitation for SCAD patients.

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Introduction

Spontaneous Coronary Artery Dissection (SCAD) is a type of myocardial infarction (MI) that presents acutely with similar characteristics as the more common atherosclerotic MI. SCAD is the cause of up to 4% of acute coronary syndrome (ACS) presentations [1]. However, in females under the age of 50, it can represent up to 35% of ACS presentations [1–3].

There is an increasing awareness of SCAD as a unique type of heart attack, and an increasing need for medical practitioners to be equipped to manage SCAD patients in the community. Thus, the aim of this chapter is to review the current definition, classification and understanding of SCAD, and highlight key similarities and differences in diagnosis (Table 4.1), management (Table 4.2) and prognosis between SCAD ACS and atherosclerotic ACS.

In addition, a schematic (Fig. 4.1) and suggested outpatient partnered management (Table 4.3) of SCAD between SCAD specialists, rehabilitation specialists, general cardiologists, and primary care providers is provided. Commonly asked questions by patients are addressed (Table 4.5), including recommendations regarding pregnancy after SCAD. Furthermore, the role of cardiac rehabilitation in this population is highlighted [4].

Definition and Classification of SCAD

The definition of SCAD is a 'spontaneous separation of the coronary artery wall, which is not iatrogenic and not related to atherosclerosis or trauma' [5]. It leads to myocardial infarction as the hematoma within the separated coronary artery wall compresses the lumen of the artery and prevents blood flow, causing ischemia.

Traditionally, SCAD was thought to result from a tear in the intima of the coronary artery, which allowed blood to flow into the intimal layer to create this false lumen that expanded and compressed the true lumen [6]. This is referred to as the "inside out" hypothesis but is now superseded largely by a modified hypothesis, comparatively named the "outside in" hypothesis which suggests that the primary incident is an intramural hematoma, rather than an intimal tear [2, 6]. The intramural hematoma is thought to be caused by spontaneous medial dissection or rupture of the vasa vasorum [2]. From there, it is believed that the underlying hemorrhage expands in either a focal, linear, or spiral fashion to compress into the lumen of the coronary artery [2, 6].

The way the coronary artery lumen is compressed is how SCAD is classified into its three types, an angiographic classification identified and labelled by the Canadian

Table 4.1 Comparison of history, physical exam, and investigations for acute coronary syndrome from atherosclerotic plaque rupture and spontaneous coronary artery dissection

Atherosclerotic ACS	SCAD ACS
History	
• Chest pain onset, timing, character • Associated features • Coronary artery disease risk factors including smoking, diabetes, hypertension, dyslipidemia, family history of premature coronary artery disease	• Chest pain onset, timing, character • Associated features • Coronary artery disease risk factors including smoking, diabetes, hypertension, dyslipidemia, family history of premature coronary artery disease • Current or recent pregnancy • Detailed obstetrics and gynecological history: – Age of menarche – Menopause onset – Pregnancies – Abortions – Multiparity (twin) pregnancies – Birth control – Hormone replacement therapy • Possible triggers (physical or emotional) • History of SCAD, hypertension or migraines • History of autoimmune disease or connective tissue disorders • Previous coronary angiograms • Family history of SCAD, vascular disorder (including fibromuscular dysplasia), autoimmune disease or connective tissue disorders
Physical examination	
• Cardiovascular exam • Respiratory exam	• Cardiovascular exam • Respiratory exam • Bruits: carotid, renal and femoral arteries • Signs of connective tissue disorders
Investigations	
• Troponin • Creatinine • Electrolytes • Fasting cholesterol • HbA1c	• Troponin • Creatinine • Electrolytes • Fasting cholesterol • HbA1c • CT angiography of head/neck and abdomen/pelvis to assess for fibromuscular dysplasia • Vascular or autoimmune work up

Table 4.2 Coronary angiogram findings, management, and course in hospital of acute coronary syndrome from atherosclerotic plaque rupture and spontaneous coronary artery dissection

Atherosclerotic ACS	SCAD ACS
Coronary angiogram	
• Locate area of plaque rupture (culprit lesion)	• Rule out acute atherosclerotic plaque disruption • Confirm SCAD diagnosis
Revascularization	
• Percutaneous coronary intervention (PCI) to stabilize culprit lesion	• Avoid revascularization if possible, unless high risk features • Natural history of SCAD is that most (96%) heal by 30 days • Higher complication rate than atherosclerotic ACS, with lower rate of PCI success
Course in hospital	
• First 24–48 h has highest arrhythmia risk	• 7% of SCAD patients treated conservatively will require intervention, generally within 5 days of initial presentation
Medications	
• Dual antiplatelet therapy • Statin • ACEI/ARB • Beta-blockers • Anti-anginal therapy	• Single antiplatelet (aspirin preferred) • Beta blocker • Blood pressure control • Antianginal therapy may be helpful to control chronic chest pain syndromes arising after SCAD

SCAD group (Fig. 4.2). Type 1 is an intimal tear with contrast staining of the false lumen, Type 2 represents a long diffuse and smooth narrowing angiographically, and Type 3 is a focal or tubular angiographic stenosis with both Type 2 and 3 resulting from an intra-mural hematoma [5].

Pathophysiology of SCAD

Recent literature has suggested conceptualizing SCAD as "SCAB" (spontaneous coronary artery "bleed") rather than "dissection", as the presence of an intimal tear is not necessary for diagnosis [7]. Increasing understanding of SCAD suggests that as the "bleed" expands between the coronary artery layers, an intimal tear can occur to decompress the intramural pressure [6]. This newer pathophysiological theory is supported by the evidence that not all SCAD cases have an intimal tear, with recent cohort studies identifying an intimal tear in only 30% of cases [2]. Further, intra-coronary imaging studies in more severe SCAD cases have shown to be associated with an intimal tear [8], which supports the "outside in" hypothesis as a unifying mechanism with the tear occurring secondarily for decompression [8, 9].

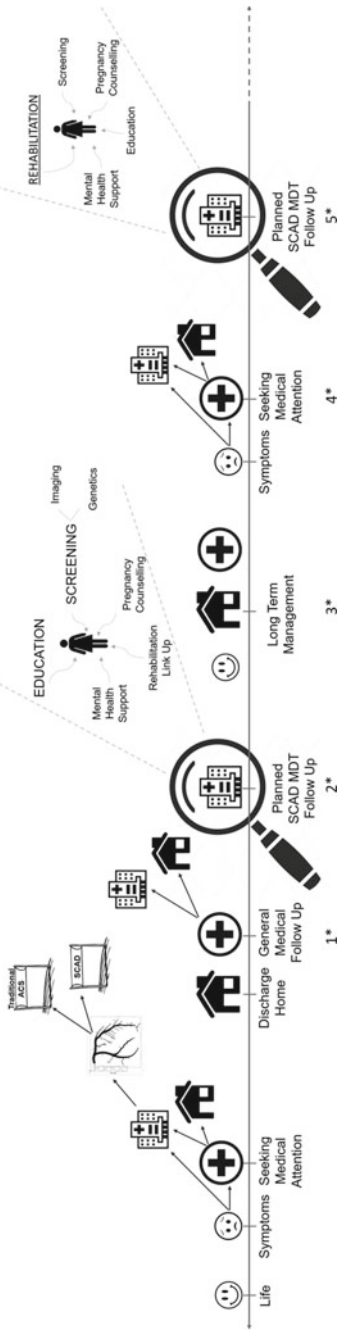

Fig. 4.1 Partnered follow up with Family Practitioner and SCAD Hospital Multi-Disciplinary Team

Table 4.3 Outpatient management following diagnosis of SCAD ACS (Correlate with Fig. 4.1)

1. Family physician follow up	
Ideally within 4–6 weeks of hospital presentation	
Aim	• Validation of the significant health event – High proportion of patients report anxiety and depression following SCAD, related to feeling extreme vulnerability from lack of understanding of SCAD and variability of information from health care providers • Pregnancy is high risk and strongly not recommended (see below) • Medical management – Hypertension management is important – Beta blockers—decreases SCAD recurrence – No role for routine repeat coronary angiography – If similar symptoms to initial presentation, need to seek medical attention, declare the prior SCAD-related MI to ED staff, have ECG and troponin measured • Lifestyle – Physical activity Regular, moderate-intensity activity is recommended Avoid heavy weightlifting or Valsalva maneuvers – Sexual activity—expected back to usual within 4 to 6 weeks. However, strongly advised against pregnancy – Food—healthy diet aimed at maintaining normal BMI, but SCAD is not a diet mediated disease • Patients must make informed decisions about their future health • Link in with SCAD support groups – Online Women@heart (www.womenheart.org) support group SCAD Patient Guide 2022 [54] (www.sunnybrook.ca/SCAD/guide) SCAD Alliance (www.scadalliance.org)
2. Planned SCAD MDT follow up I	
Ideally within 8–12 weeks of hospital presentation	
Aim	• Consultation reviewing SCAD and understanding of its mechanism • One-time cross-sectional imaging from head to pelvis with CT angiography looking for: – Fibromuscular dysplasia (tortuosity, aneurysms, stenoses, dissections, contour irregularities (beading)) • Pregnancy is high risk and is not recommended – Due to the possibility of severe and potentially life-threatening SCAD during pregnancy, and recurrence rates of 17% for women with peripartum SCAD, pregnancy after SCAD-related MI is not recommended – In the event of pregnancy—patients must seek involvement of SCAD expert and high-risk obstetrics team prior to pregnancy or as early as possible after pregnancy to consider their options • Further cardiac investigations if indicated, which may include – Echocardiogram; – Cardiac MRI (rule out differentials) – Coronary CT angiography or repeat coronary angiography—rarely required • Review of medication • Review of associated conditions • Review of psychosocial state • Confirm referral/attendance at cardiac rehab

(continued)

Table 4.3 (continued)

3. Long term management	
Aim	• Risk stratifying your patient to likelihood of recurrence of SCAD, higher likelihood in those with – Migraine headaches – Poorly controlled hypertension – Fibromuscular dysplasia diagnosis – Known coronary artery tortuosity (from initial angiogram) Current literature suggests that receiving coronary stents is not associated with prevention of recurrent SCAD in future. Further, if SCAD recurs, it tends to affect different coronary arteries from that which caused the initial event
4. Seeking medical attention with unplanned chest pain presentation	
Higher concern if within 5–6 days of initial presentation	
Aim	Determine need for hospital management • Immediate hospital management if – Hemodynamically unstable – Acute ongoing chest pain – Dynamic ECG changes, dynamic rise and fall of troponin • Likely community management if: – Hemodynamically stable – Symptoms resolved – Patient appears well – No troponin elevation, or falling troponin compared with recent hospitalization
5. Planned SCAD MDT follow up II	
Aim	• SCAD education – Increasing education as more research on SCAD conducted and medical understanding increases • SCAD rehabilitation, involving: – Graduated exercise regimen • SCAD psychosocial support, involving: – Cognitive behavioral therapy (CBT) as needed – Peer support (Women@heart: www.womenheart.org) • Education regarding SCAD and pregnancy

Epidemiology of SCAD

The mean age of SCAD patients is between 44 and 52 years, and it is uncommon in the extremes of age (under 25 years or over 80 years) [5, 10]. The female predominance is now well established in the literature, with a large meta-analysis of 2172 patients revealing 84% of SCAD patients as female [6].

There has been a temporal increase in the incidence of SCAD, with a rise in awareness paralleling a rise in diagnosis, particularly since 2012 [2]. Although it is likely still under-recognized at present, SCAD is believed to be the cause of up to 4% of ACS presentations [3]. Further, in administrative databases, SCAD represents 15–20% of myocardial infarctions during pregnancy or peripartum, a particularly high-risk period for known SCAD patients [1, 10, 11].

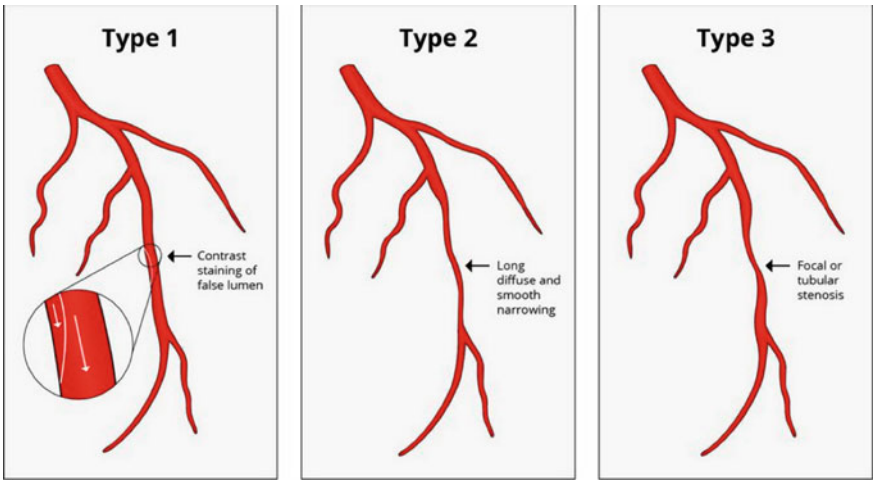

Fig. 4.2 Classification of SCAD**.** *Figure from Saw, 2016, contemporary review of spontaneous coronary dissection* [5]

Recent data from the Canadian SCAD registry demonstrated that recurrence rates for SCAD are surprisingly low, 2.4% at 3 years [12]. Further, the 30-day mortality rate is 0.1% [13] and 3-year mortality rate is 1% [14] for SCAD patients.

Risk Factors for SCAD

The most notable risk factor for poor outcome in SCAD is pregnancy and the post-partum state in a patient with prior SCAD. Hypertension also is a risk factor for SCAD-related myocardial infarction during pregnancy [15]. Amongst the SCAD patient population, 15–20% of patients are pregnant or post-partum at the time of diagnosis [1, 16]. However, the exact mechanism of association between hormones and SCAD is not yet understood, and the data is conflicting, as there does not appear to be a clear association between oral contraceptives or hormone replacement therapy and SCAD [16].

Secondly, hypertension has a high prevalence (45%) in SCAD patients [15]. There is a plausible explanation mechanistically, as hypertension can lead to chronic pathological changes in the arterial wall via increasing arterial wall stress, endothelial damage, triggering smooth muscle cell proliferation and breakdown of elastin fibers, that ultimately make the vessel more vulnerable to SCAD [15]. Hypertension is also independently associated with a higher risk of in-hospital mortality (OR 2.19, CI 1.86, 2.58) [17]. Furthermore, in a prospective Spanish registry, hypertensive SCAD patients had higher risk of increased severity of SCAD lesion (15 versus 7%, p >

0.05), higher risk of procedural complications (65 versus 41%, $p < 0.05$) and lower likelihood of procedural success (65 versus 88%, $p < 0.05$) [15, 18].

In addition, there should be clinical suspicion for SCAD in the presence of risk factors such as fibromuscular dysplasia (FMD). Between 15 and 70% of SCAD patients are found to have FMD with the wide range explained by variable screening protocol [1, 3, 6, 19]. Further risk factors may include connective tissue disorders, autoimmune and inflammatory disorders [1, 6], as well as microvascular dysfunction [15, 20], however there is not enough data to establish a clear association between these entities and SCAD at present. The role of genetics in the mechanism of SCAD is also not yet fully understood. A recent study noted a higher prevalence of fibrillin 1 (FBN1) in the plasma of SCAD patients compared with non-SCAD ACS patients [21]. This medium sized ($n = 70$ in each arm) study generated interest due to the mechanistic plausibility as fibrillin 1 is a component of the elastic tissue found in the media of coronary arteries [21]. Presently, there are no specific genetic screening tests for SCAD or fibromuscular dysplasia, but this is an active area of research [22].

Clinical Presentation of SCAD

As a type of ACS, SCAD presents acutely with similar characteristics as atherosclerotic myocardial infarction. Although ACS presentations in young and middle-aged females with no atherosclerosis risk factors should lead to a high suspicion for SCAD, it must be emphasized that ACS by atherosclerotic plaque rupture is significantly more common than SCAD (50 versus 4%) [23]. Thus, atherosclerotic ACS must be ruled out by coronary angiography before the diagnosis of SCAD can be considered in any clinical presentation [10].

Similar to atherosclerotic ACS, most patients with SCAD (85–96%) experience chest pain or discomfort as their primary presenting complaint [10]. However, a small proportion of SCAD patients may have atypical symptoms including dyspnea (20%), back pain (14%), diaphoresis (21%), nausea and vomiting (24%) and presyncope (9%). Clinically, approximately 20–50% of SCAD patients present as STEMI, and up to 5% present with ventricular arrhythmia, and 2% with cardiogenic shock [10].

In addition, SCAD should be suspected in patients with associated extreme physical or emotional trigger, which is notable in 40% and 24% of SCAD presentations, respectively [9]. The postulated mechanism is a hyper-catecholaminergic state, which along with Valsalva-like maneuvers in physical or emotional states can damage the vasculature and arterial wall with shear force [6]. In the recently published study by the Canadian SCAD group, an isometric physical trigger was more commonly observed in men (40.2 versus 24.0%, $p = 0.007$), whilst emotional stress triggers have been found to be more likely in women (35 versus 60%, $p = 0.001$) [9, 24].

What to Do if You Suspect Your Patient Has SCAD

A detailed history and physical examination are key to timely and accurate diagnosis of SCAD (Table 4.1). When there is a suspicion of SCAD, the assessment needs to include questions regarding previous SCAD, pregnancy, menstrual cycle, migraines, physical (particularly isometric) or emotional triggers, known FMD and vascular disorders. Similarly, the physical examination should extend beyond the cardiovascular exam to include auscultating for renal bruits and assessing for signs of connective tissue disorders.

Investigation and management are initially similar for suspected SCAD patients and atherosclerotic ACS patients (Table 4.2). This includes serial electrocardiograms (ECG) and troponin levels. Following that, coronary angiography is vital as it can visualize plaque rupture to identify traditional atherosclerotic ACS, or rule out plaque rupture to confirm the suspected diagnosis of SCAD [10].

We know that there are also ACS presentations that are neither plaque-rupture atherosclerotic ACS nor SCAD, and can be caused by myocardial infarction with no obstructive coronary arteries (MINOCA), coronary vasospasm or microvascular dysfunction. Presentations can also be ACS-mimics such as myocarditis, myopericarditis, takotsubo cardiomyopathy, or other non-cardiac diagnoses including pulmonary emboli or aortic dissection.

Having considered the wider differential for acute chest pain, the general principle is to treat the patient as atherosclerotic ACS until SCAD or an alternate differential is diagnosed on coronary angiography [15]. If SCAD is the true diagnosis, the course of management significantly changes, including avoiding invasive percutaneous coronary intervention (PCI) if possible [18]. However, PCI may still be considered in the presence of clinical high-risk features, including ongoing ischemia/chest pain, cardiogenic shock, sustained ventricular arrhythmia and left main dissection [25].

Further, coronary angiography assists in determining the type of SCAD and the amount of myocardium affected, which can also influence management and follow up frequency and strategy [26]. It is important to explain to the patient the risks of coronary angiography for informed consent, including vascular damage, stroke, myocardial infarction, arrhythmia, contrast risk and focusing specifically on radiation risk for young female patients. Also, the possibility of pregnancy must be assessed, and risks must be discussed.

Moreover, SCAD patients are expected to be admitted in hospital for a longer duration of time (5–7 days) than atherosclerotic ACS patients, as studies have shown that SCAD patients who deteriorate do so usually within 5 days of presentation, compared to atherosclerotic ACS where the highest risk period is the first 48 h [27].

Conservative Versus Invasive Management

Management for revascularization differs between SCAD and atherosclerotic ACS patients. It is well established that if the clinical situation allows, conservative management without PCI is the preferred treatment strategy for SCAD patients, whereas for an atherosclerotic plaque rupture, PCI is recommended. This is explained by the natural history of SCAD, whereby 95% of SCAD are healed spontaneously by 30 days [25]. However, 5–7% of patients who were initially treated conservatively will require PCI in the same presentation, most often due to cardiogenic shock, ongoing ischemic chest pain, or proximal coronary artery involvement [8, 25].

Intervention is considered only for select clinical situations in SCAD because the presence of dissection and intramural hematoma adds to the complexity of the procedure, resulting in higher complication rates [2]. The success rate of PCI for SCAD is 50–70% in large cohort studies, which is much lower than that for the atherosclerotic ACS PCI population [1, 28, 29]. Further, although this may be impacted by a selection bias of the more severe SCAD patients receiving intervention, PCI for SCAD has not been independently associated with reduced infarct size, nor with reduced in-hospital MACE (Major adverse cardiovascular events) compared to conservatively managed SCAD groups [27, 30].

Medical Management

The main debate in the acute medical management of SCAD ACS has been regarding single or dual antiplatelet therapy use. Currently, there are no randomized clinical data or established protocol [31], but there is expert consensus opinion to offer guidance. The most recent literature favors single antiplatelet therapy (SAPT) for SCAD ACS over dual antiplatelet therapy (DAPT) with the multicenter DIssezioni Spontanee COronariche (DISCO) registry from Italy and Spain ($n = 199$) showing patients on DAPT ($n = 132$, 66%) experienced significantly higher MACE (defined as all-cause death, non-fatal MI or unplanned PCI; 18.9 versus 6.0%. HR 2.62, 95% CI 1.22–5.61, $p = 0.013$) than those treated with SAPT ($n = 67$, 34%) [32]. This result was driven mainly by non-fatal MI (15.2 versus 3.0%, $p = 0.009$) and unplanned PCI (12.1 versus 1.5%, $p = 0.001$) [32]. The pathophysiological rationale behind this is increased bleeding and intramural hemorrhage due to DAPT, with no added benefit as there is no acute plaque rupture or disturbance. Similarly, the consensus is that anticoagulation is not necessary for SCAD ACS [1, 2, 33].

The key medication for the management of SCAD long term are beta blockers. A single center observational study has shown a significant decrease of SCAD recurrence with the use of beta blockers, by over 50%, over 3 years [33]. The second step of optimal medical management of SCAD is good blood pressure control, aiming for SBP < 130 mmHg. Angiotensin Converting Enzyme-Inhibitors (ACE-I) or Angiotensin Receptor Blockers (ARBs) are used to treat hypertension, which

is a clear risk factor for both SCAD ACS and its recurrence, and are also indicated for the management of left ventricular dysfunction after SCAD-related myocardial infarction. If more blood pressure control is necessary, calcium channel blockers or diuretics may also be used as secondary agents, in line with treatment guidelines for essential hypertension [34].

Unlike atherosclerotic ACS, there is no evidence for the routine use of statins in SCAD [31]. Cholesterol-laden plaque is not implicated in the pathophysiology of SCAD ACS, while it is central to the pathophysiology of traditional atherosclerotic-myocardial infarction. This is a challenging concept for most cardiologists and family physicians to accept, as the use of statin therapy after ACS is firmly entrenched in the practice patterns of most physicians. Similarly, anti-anginal medications (nitrates, and calcium channel blockers) may be very useful for symptom control for chest pain syndromes after SCAD, but are not prescribed routinely [2].

Outpatient Management of SCAD Patients

The optimal outpatient management of SCAD patients is continually being developed as SCAD becomes more understood. Currently, as a lesser-known form of ACS with minimal public awareness or education, many SCAD patients experience social isolation, anxiety and depression during this vulnerable period in their recovery [4].

Indeed, good quality clinical research on outpatient management of SCAD patients are lacking, with many current recommendations being derived from the larger vessel (aortic) dissection literature whilst direct experience with SCAD patients is still building. Thus, the partnered management of patients between SCAD specialists, general cardiologists, cardiac rehabilitation specialists and primary care providers is important for the communication of accurate information (Table 4.5), education of effective management strategies, and screening of associated pathologies [35]. Such management partnerships should be identified and linked in with the SCAD patient prior to hospital discharge.

This schematic (Fig. 4.1) is a timeline from the point of SCAD ACS presentation to diagnosis and management in the hospital, followed by discharge home and the subsequent outpatient avenues of follow up (Table 4.3) with both the primary care provider (Time points: 1, 3, 4), and the planned SCAD multi-disciplinary team (MDT).

The first follow up with the family physician (Point 1* in Fig. 4.1 and Table 4.3) after discharge from a SCAD admission to hospital is recommended to occur within 4–6 weeks. One of the key message that needs to be reiterated to young females of child-bearing age is that after SCAD ACS, pregnancy is not recommended due to the high risk of severe and potentially life-threatening recurrence of SCAD during pregnancy or post-partum [16, 36]. Thus, in the event of pregnancy despite such warning, patients must seek involvement of SCAD expert and the high-risk obstetrics team as early as possible to consider their options. Seeking expert opinion prior to conception is most ideal if pregnancy is desired despite awareness of its high risk.

Regarding exertion, SCAD patients are often advised to avoid high intensity physical exertion and Valsalva maneuvers, which involves not lifting anything heavier than 30lbs for women, or 50lbs for men. However, this should not preclude patients from regaining an active lifestyle with regular moderate activity post SCAD. Participation in cardiac rehabilitation as early as possible is vital in facilitating this.

Between 8 and 12 weeks after hospital discharge, a SCAD MDT follow up (Point 2* in Fig. 4.1) is ideal for further assessment and investigations for associated disorders. The most important and well documented association is FMD, and all patients who have experienced SCAD must have an initial screening test for FMD via a head-to-pelvis computed tomographic (CT) angiography looking for aneurysm, tortuosity, dissection, irregularity, or stenosis of arterial beds. This may have already been done during the inpatient stay if a CT angiography was available.

During the late follow up period, an unplanned chest pain presentation (Point 4* in Fig. 4.1) may occur. These need to be approached in a similar way to a new chest pain presentation, without presumption that it is related to SCAD as recurrence rates of SCAD are very low (2.4% over 3 years) [37, 38]. Thereby, hemodynamic stability, acute onset of symptoms and dynamic ECG changes and troponin levels are used to determine if the patient is best for in-hospital or community management. Notably, women are significantly more likely than men to be re-admitted to hospital for chest pain following prior SCAD (8 versus 1%, $p = 0.001$) [24].

Further, these outpatient scheduled reviews or unscheduled chest pain presentations are opportune to review medications and ensure patients are taking beta blockers long term following their SCAD ACS episode, assess their psychosocial state, and ensure referral to outpatient cardiac rehabilitation [26].

Longer term, after two to three years of consistent follow up by the SCAD MDT and a stable clinical state, shared decision making is advised to individualize the best method for ongoing patient access to SCAD resources and healthcare. Anecdotally, most patients and family physicians have felt comfortable at this stage to take ownership of the follow up long term, given the established relationship with the SCAD MDT and ability to re-refer if concerns arise.

Cardiac Rehabilitation

Cardiovascular rehabilitation (CR) is a comprehensive secondary prevention program that includes patient education, nutritional counseling, exercise training and psychosocial interventions [39, 40]. Participation has been associated with up to a 50% reduction in morbidity and mortality, as well as improved quality of life following a cardiac event such as MI or cardiac procedure [41].

Unfortunately, less than 30% of eligible patients participate in CR [42] and women in particular, are less likely to be referred and participate in programming [43]. Younger age at diagnosis, lack of referral, caregiving/family responsibilities, transportation, scheduling and logistical difficulties with attending an onsite program have been identified as predominant barriers for enrollment in CR [26, 44, 45].

Despite the significant benefits associated with CR program completion, women diagnosed with SCAD are significantly under-represented in CR programs across Canada [12, 46]. Fear, anxiety and hesitancy regarding physical activity after SCAD are common and often lead patients to curtail or avoid physical activity altogether [11]. CR programs are recommended to safely support the resumption of moderate intensity physical activity after SCAD-related events [11, 47, 48]. Recommendations for physical activity should be tailored to the individual and may be best determined and guided during participation in CR [47, 48].

Cardiac Rehabilitation and SCAD Patients

Referral to CR is considered the standard of care for all patients who have experienced a cardiac event or procedure, including those with a SCAD diagnosis [40].

Recent North American studies have shown improvements in aerobic capacity, body composition and mental health with no reported adverse events in SCAD patients completing CR [46, 48]. Structured rehabilitation (Table 4.4), particularly multi-dimensional interventions that offer comprehensive components (e.g. exercise, psycho-educational support, mindful living sessions, nutrition counselling, peer support networks) have produced both physical and emotional benefits [49].

Krittanawong et al. [49] examined the usefulness of CR in a large cohort of SCAD patients (48/50 US states, Canada, Europe, Australia and New Zealand) enrolled in the Mayo Clinic SCAD registry (n = 354). In this cohort, 66% of patients participated in > 10 CR sessions while 32% chose not to participate. A lack of recommendation to CR by a health care provider was cited as the primary reason for non-participation. An inherent reluctance for clinician referral may be attributed to the relative unfamiliarity with SCAD, a lack of perceived benefit in this population due to the younger age without traditional risk factors, and concerns that exercise training may prompt a recurrent SCAD event.

Wagers et al. [50] further reinforced the positive experiences and safety outcomes for SCAD survivors (n = 409) participating in a CR program. However, a focus on the 'typical' or traditional CR program was deemed by patients as "not a good fit" considering the patient's younger age, gender and/or prior activity levels. Some participants reported feelings of isolation during CR due to lack of younger counterparts to whom they could relate, while others felt their program was geared to the "standard heart attack" patient. Baechler et al. [26] interviewed 38 patients diagnosed with SCAD and observed that frustration with the lack of mental health resources was mentioned among the most common reported themes. Patients described that an ideal CR program should be specific for patients with SCAD and would include mental health support [26]. The importance of psychosocial support for SCAD survivors has been highlighted in several studies [51, 52] and reinforces that it should be accessible to every patient attending CR [39, 40], notably to women [53].

Neubeck et al. [52] performed an extensive systematic review of 28 studies and analyzed the physical and psychosocial recovery following discharge from hospital

Table 4.4 Recommendations for cardiac rehabilitation (CR) programs treating SCAD survivors

CR program duration	6 months
Cardiovascular risk factors	• Traditional and non-traditional risk factors for cardiovascular disease should be assessed and risk profile shared with patients • CR programs should follow the current guidelines on management of blood glucose, hypertension, body weight, physical activity, psychosocial wellbeing, sleep, healthy diet, and smoking cessation • Note that SCAD ACS is not considered a risk factor to initiate statin therapy. SCAD is not a statin-indicated condition • Statin therapy should be considered when LDL-cholesterol is very high, > 5.0 mmol/L or as recommended by the current guidelines for primary prevention of cardiovascular disease (if there is no established atherosclerosis)
Exercise structure	• One-hour weekly exercise class consisting of 15 min warm-up, 30 min cardiovascular exercise on aerobic machines, and 15 min cool-down
Frequency	• 30–40 min of moderate intensity physical activity 5–7 days/week (150 min/week)
Target exercise heart rate	• 50–70% of heart rate reserve based on the entrance exercise treadmill test
Blood pressure limit	• < 130/80 mm Hg
Target perceived exertion rate	• “Moderate” to “somewhat difficult”
Resistance training	• Focus on a proper breathing and lifting technique avoiding the Valsalva maneuver/straining
Weightlifting	• 2- to 12-pound free weights to increase muscle strength, starting with lighter weights and progressing with strength gain • Avoid lifting weights > 30 pounds for women and > 50 pounds for men
Educational sessions	• 20 min educational session per week on risk factors, and treatment of heart disease and SCAD, and stress management, emphasizing women’s heart disease when appropriate
Mental health	• Counselling, mindful living sessions, and peer-support from other SCAD survivors • Peer Support from patients with lived experience has been identified as a key component in recovery following a cardiac event such as SCAD
Medications	• If medication changes (eg: beta-blocker dose), consider repeating cardiopulmonary test to support optimizations of exercise prescription
Health care team	• Physician, Kinesiologist, Physiotherapist, Dietitian, Psychologist, Social Worker, Registered Nurse
Recommendation to patients	• Stop or slow down if becoming extremely exhausted or feeling uncomfortable, or if symptoms arise, including but not limited to chest discomfort, chest pain, dyspnea, or dizziness

Table 4.5 What to tell my SCAD patients?

Have I had a heart attack?
Yes, but a different kind of heart attack. A heart attack is when there is no flow through one of the heart arteries to the heart muscle, causing damage to the heart muscle. Most commonly, this occurs from cholesterol plaque buildup in the lining of a heart artery which ruptures and blocks the artery (and thus prevents the blood from flowing to the heart muscle) However, with SCAD, the blood flow blocks from a bleed that occurs within the layers of the heart artery. This causes compression of the heart artery from the outside, leading to reduced blood flow or in some cases, a tear in the artery lining
If I have had a heart attack, why did I not get a stent?
Some people with SCAD require a stent because the compression of the heart artery from the outside is very severe. However, it is preferrable to avoid a stent as it can lead to other complications such as extension of the tear in the artery or making the amount of artery compression more extensive. You did not have a stent inserted because you were still able to get enough blood flow to your heart muscle As the underlying problem is not a plaque within the heart artery, but a compression from the outside, all current research point to the best outcomes for SCAD if you can wait for the "bruise" in the wall of the heart artery to slowly dissolve itself over time
Will I have SCAD again?
A small proportion of people do get SCAD again. Understandably, this can be distressing Having had SCAD however, you are in a better position to know what it feels like and seek medical attention early. Thankfully, most people do not get SCAD recurrence and most SCAD patients recover well and safely
What do I need to avoid getting SCAD again?
The main thing to do is ensure your blood pressure is under good control, through lifestyle changes and medication. Stay on the medications prescribed by your doctor, which may include beta blockers. They aim to reduce the pressure stress on your heart arteries and have been shown to lower the chance of SCAD recurrence It is also important to avoid known physical triggers such as extreme heavy lifting that involve sudden Valsalva (bearing down) maneuvers
Why am I getting so many mixed messages?
Again, this is understandably confusing. Although the knowledge of SCAD is increasing over the years, particularly compared to regular heart attacks it is a relatively uncommon diagnosis. Thus, the understanding of it amongst the general community and the medical community is much less than for a traditional heart attack
Can I get pregnant?
Informed decision making around pregnancy is very important for SCAD patients. Pregnancy cannot be recommended because of the increased risk for another SCAD related heart attack which may be much more severe during pregnancy. Continuation of pregnancy, or the decision to terminate pregnancy must be a very carefully considered decision along with a specialized multidisciplinary team. This is because SCAD is the most common pregnancy-related heart attack and is thought to be related to the hormone changes that occur in pregnancy, particularly in the early post-partum period

in 4167 patients (93.5% female) with SCAD. They observed lack of specific guidance about physical activity and high levels of psychosocial distress. These findings suggest ideal programs with tailored exercise and psycho-educational components including peer support networks require further examination to test feasibility and effectiveness specific to the unique needs of these patients. Moreover, it is imperative that care providers are aware of the safety and associated benefits of CR participation to improve the recovery trajectory for this understudied population.

Physical Activity After SCAD

Fear and hesitancy regarding physical activity after SCAD are common among SCAD survivors and clinicians despite a lack of evidence for protection from recurrent SCAD [11]. Additionally, imposing physical restrictions to young individuals could lead to lifelong impact possibly promoting sedentarism, weight gain, and psychosocial issues. In this realm, CR programs can contribute substantially to overall physical and mental health [11].

As outlined previously, CR has demonstrated overall benefit and safety [46, 48–50]; therefore, pursuing regular, moderate exercise likely outweighs the theoretical risks of recurrent SCAD [11, 47, 48]. It is important to note that SCAD survivors, especially those who experienced recurrent SCAD or who have noncoronary aneurysms or dissections should avoid extreme endurance training, exercising to exhaustion, elite competitive sports, or vigorous exertion in extremes of ambient temperature [11, 47, 48].

The Vancouver General Hospital (VGH) SCAD-CR protocol has been adopted worldwide and provides safe parameters for patients initiating CR [48]. Each patient should have an individualized exercise prescription, considering clinical parameters, physical exercise prior to SCAD and personal goals. These considerations do not substantially differ from non-SCAD patients however, it is suggested to start physical activity at a lower level, as recommended by Chou et al. [48], then gradual progression should be targeted (Table 4.4). Further, SCAD survivors should avoid lifting or carrying heavy objects that require straining or prolonged Valsalva [11, 47, 48]. Nevertheless, there is no evidence that heavier loads with proper technique (no straining or Valsalva) are harmful [47].

The length of time between a SCAD event and CR initiation should be considered, and empirically exercise prescription could be more conservative in the first months post SCAD as suggested in Table 4.4, and less restrictive after 6–12 months. The presence of FMD can also represent an additional challenge. Patients with carotid or vertebral artery dissections should avoid resistance training including body weight exercises such as push-ups and sit-ups during the first 8–12 weeks after the acute dissection after which the recommendations would be similar as for SCAD [47].

Future Directions and Opportunities

The evolving understanding of SCAD over the last ten years is promising to translate into partnered care and better outcomes for SCAD ACS patients. More robust data have clarified prior assumptions, and management strategies have become more unified globally. The concern for pregnancy following a SCAD ACS episode continues, particularly as recurrence in this scenario is more likely to be severe.

There are numerous future opportunities that need to be addressed in order to better understand the presentation, diagnosis, early recognition and treatment as well as long term management of patients diagnosed with SCAD.

Cardiac rehabilitation is safe and feasible, early studies have shown the beneficial effects on clinical cardiovascular parameters and mental health in SCAD survivors [46, 49, 50]. Future studies need to specifically examine the clinical experiences and preferences of SCAD patients in order to develop a better understanding of alternative, flexible program models, the integration of peer support networks and how best to tailor CR programming to meet the patient's psychosocial, physiological and educational needs.

It is crucial that care providers are educated and understand the differences between SCAD and atherosclerotic diseases, and how best to mitigate barriers to SCAD care. Consideration of family systems and roles as well as involvement of family members in SCAD education may help alleviate patient and family anxiety which may subsequently optimize recovery transitions.

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