Case 1: A 34-year-old gravida 2, para 2 woman was admitted with anterior ST-segment–elevation myocardial infarction 10 days postpartum. Coronary angiography showed left main (LM) dissection with occlusion of the proximal left anterior descending (LAD) artery (Figure 1A), and her ejection fraction was 35%. Bare metal stents were successfully implanted from the LM into the proximal LAD and diagonal arteries. Antegrade extension of dissection into the circumflex was left untreated because her ischemia resolved. However, repeat angiography at 5 months with optical coherence tomography showed in-stent restenosis in the LM, stent-strut malapposition in the proximal LAD (Figure 1B), and residual circumflex dissection, prompting subsequent coronary artery bypass grafting (CABG).

Case 2: A 40-year-old woman (gravida 2, para 2) presented with non–ST-segment–elevation myocardial infarction 10 days after elective cesarean section. Coronary angiography showed proximal LAD dissection with 80% stenosis (Figure 1D), and her ejection fraction was 40%. She was treated conservatively because her chest pain resolved. Repeat angiography at 5 days showed improvement of stenosis to 50% (Figure 1E). Computed tomography angiography at 14 months showed almost complete normalization of the proximal LAD. She was found incidentally to have fibromuscular dysplasia of her external iliac artery (Figure 1F).

Introduction
Pregnancy-related (P-) spontaneous coronary artery dissection (SCAD) is a rare and potentially lethal complication of pregnancy. It is estimated that 1 in 16,000 pregnancies is complicated by acute myocardial infarctions in the United States; up to one fourth of these are due to P-SCAD. Although the true prevalence of overall SCAD is unknown and older reports suggested that pregnancy caused a substantial portion of SCAD, contemporary series estimated that P-SCAD accounted for only <5% of SCAD cases.

SCAD is defined as a separation within the arterial wall by intramural hematoma, which can occur by an intimal rupture initiating medial dissection or more commonly by a spontaneous intramedial hemorrhage such as that resulting from disruption of the vasa vasorum (Figure 2). P-SCAD can occur during pregnancy (as early as 2 weeks after

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conception) or postpartum, defined as within 6 weeks of delivery by the World Health Organization. The term peripartum SCAD has variable and limited definitions, ranging from the third trimester of pregnancy to 3 to 5 months postpartum. We have also observed late P-SCAD several months postpartum, even as late as 11 and 16 months postpartum, especially in women still breastfeeding. Thus, the term P-SCAD is preferred for broader inclusion and may be categorized as antepartum, early postpartum (within 6 weeks of delivery), late postpartum (6 weeks to 12 months), and very late postpartum (12–24 months).

Multiple predisposing causes of P-SCAD have been described. Estrogen and progesterone surges during pregnancy (especially recurrent exposures with multiparity) can cause structural changes with recurrent and chronic accumulation of medial degeneration such as that from decreased collagen synthesis and increased media mucopolysaccharide content, causing weakening of the tunica media. Histopathological features include fragmented and disorganized elastic and collagen fibers, microcystic mucinous pools, cystic medial necrosis, and inflammatory infiltration at arterial dissection planes. In particular, periadventitial eosinophilic infiltration was observed, but it is unclear if this was reactive to or causative of SCAD. Aside from pregnancy-related structural changes, fibromuscular dysplasia was recently discovered to be strongly associated with SCAD and was reported with P-SCAD. Other systemic predisposing causes such as connective tissue disorders (eg, Ehlers-Danlos type IV, Marfan syndrome) and inflammatory conditions (eg, systemic lupus erythematosus, ulcerative colitis) may also be associated with P-SCAD but are infrequently linked. The physiological changes during pregnancy, with >50% increase in cardiac output, and the hemodynamic effects of labor-intensifying vascular shear stresses can also precipitate P-SCAD when superimposed on arterial structural changes related to pregnancy or other predisposing cases. The association of breastfeeding with late and very late postpartum P-SCAD is intriguing and may suggest that hormonal changes

Figure 1. A, Case 1: left main dissection (+) with smooth stenosis extending into the left anterior descending coronary artery (LAD), which is occluded proximally (*). B, Optical coherence tomography at follow-up showing severe stent strut malapposition in the proximal LAD. C, Case 2: extensive dissection with multiple radiolucent lumen (*) originating from the ostial LAD and occluded in the midsegment (arrow). D, Case 3: proximal LAD dissection with smooth 80% stenosis, which improved 5 days later to ≈50% stenosis (E) with contrast hang-up in the arterial wall (arrow). F, Fibromuscular dysplasia changes of the right external iliac artery in case 3.
Clinical Presentation and Diagnosis

The mean presenting age with P-SCAD was 33 years in the 3 largest medical literature review series, with timing at a mean of 23 to 24 days postpartum or 32 weeks’ gestation. The vast majority (>85%) were multiparous, with a mean parity of 2.4 to 2.8 and gravidity of 2.7 to 3.1.8,10,14 Clinical presentations included chest pain, dyspnea, acute myocardial infarction, congestive heart failure, ventricular arrhythmia (including sudden cardiac death), and cardiogenic shock. Such presentations in pregnant or postpartum women should prompt early invasive coronary angiography to rule out P-SCAD, with precautions to minimize radiation exposure in antepartum women.

The gold standard imaging is coronary angiography for SCAD; computed tomography angiography has inadequate spatial resolution to visualize SCAD in smaller coronary arteries, especially those <2 mm in diameter.6 Given the possibility of iatrogenic catheter-induced dissection in patients with arterial fragility prone to SCAD, cautious and meticulous angiographic techniques are warranted with careful attention to pressure tracing, avoidance of deep intubation (especially with the radial approach in which iatrogenic LM dissections were reported with SCAD),3 gentle contrast injections (to minimize propagation of dissection), and consideration of initial nonselective injections to visualize the LM. Despite these measures, propagation of dissection may still occur, and prompt recognition and management are imperative. The angiographic appearance, categorization, and algorithm for SCAD diagnosis have been described previously.15 The stereotypical appearance of contrast arterial wall stains (type 1 SCAD) is present in fewer than one third of cases; instead, the most common angiographic appearance is a long diffuse stenosis (type 2 SCAD) without intimal disruption, as depicted in our cases. Intravascular ultrasound or optical coherence tomography is useful to confirm SCAD in nonstereotypical cases. Vascular calcification and other changes typical of atherosclerosis are

Figure 2. A, Normal coronary arterial wall. B, Spontaneous coronary artery dissection (SCAD) with intramural hematoma in the arterial wall without intimal rupture, which may be related to vasa vasorum rupture. Predisposing histopathological arterial wall abnormalities of periadventitial inflammation, cystic medial necrosis, fibromuscular dysplasia, and medial degeneration are depicted. C, SCAD with intimal rupture depicted as the cause of dissection and intramural hematoma.
usually absent. Patients with P-SCAD also have a tendency for proximal coronary involvement (including the LM and LAD), multivessel dissections, and worse left ventricular ejection fraction compared with patients with non-P-SCAD.9

Management
An algorithm to guide management, including percutaneous coronary intervention (PCI) and surgical interventions (eg, CABG, extracorporeal membrane oxygenation or left ventricular assist device), is described in Figure 3. Conservative therapy is preferred in the management of stable SCAD patients,6,16 especially because the vast majority of dissected segments heal spontaneously (We reported 79 of 79 cases of healing on repeat angiograms after 4 weeks).3 The primary factors necessitating revascularization are the presence of ongoing ischemia/infarction, hemodynamic instability, and involvement of the LM, which, unfortunately, are frequently encountered with P-SCAD.

PCI is typically the first-line revascularization strategy for SCAD for feasible anatomy except for LM and ostial LAD dissection, in which case CABG is generally preferred. Unfortunately, PCI for SCAD is fraught with challenges, with technical success rates of ≈65%.3,17 Even if PCI is technically successful, extension of dissections occurs in 25% to 60% of cases, and long-term durable results were achieved in only ≈30%.3,17 Wiring into the true lumen can be challenging, especially in the presence of intimal disruption. Confirmation of true lumen position may be achieved with intravascular ultrasound or optical coherence tomography, before angioplasty or stenting. Strategies to minimize hematoma extension include implanting a very long stent that extends 5 to 10 mm on both sides of the dissection and stenting the distal edge first (to limit apical vessel extension), followed by the proximal edge and then the middle section.6,18 Optical coherence tomography or intravascular ultrasound is also useful to achieve better short- and long-term results by ensuring adequate dissection coverage and appropriate sizing. Drug-eluting stents are typically used, given the extensive lengths required and the risks of subsequent restenosis observed.3 In addition, because intramural hematoma spontaneously resorbs over weeks, subacute and late malapposition (as shown in case 1) with the risk of stent thrombosis is a potential concern. Thus, drug-eluting bioabsorbable stents may have possible advantages.

CABG is typically reserved for patients with LM dissection or patients in whom PCI is not feasible or is unsuccessful. Similar to PCI, there are technical challenges with CABG, including isolating suitable nondissected graft insertion sites or surgically approximating vessel layers at the site of graft insertion. Long-term durability with surgical grafts for SCAD was reported to be poor. In the Mayo Clinic series, 11 of 15 grafts were occluded at the long-term follow-up,17 probably reflecting the natural history of spontaneous healing of dissected segments, resulting in eventual graft failure from competitive flow. However, the suboptimal long-term graft patency should not detract from choosing CABG in patients in

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**Figure 3.** Recommended management algorithm for pregnancy-related (P-) spontaneous coronary artery dissection (SCAD). CABG indicates coronary artery bypass graft; Circ, circumflex; CP, chest pain; ECMO, extracorporeal membrane oxygenation; IABP, intra-aortic balloon pump; ICD, implantable cardioverter-defibrillator; LAD, left anterior descending artery; LM, left main; LVAD, left ventricular assist device; Med tx, medical treatment; PCI, percutaneous coronary intervention; VF, ventricular fibrillation; and VT, ventricular tachycardia.
whom PCI is not feasible because, in many life-threatening cases, it is the only means of establishing coronary flow and salvaging ischemic myocardium acutely. In these instances, the surgical intervention (often with hemodynamic support) enabled short-term survival of these patients and myocardial preservation.

Mechanical support with intra-aortic balloon pump is often required for hemodynamically unstable patients. The use of extracorporeal membrane oxygenation and a left ventricular assist device should be considered for patients with persistent cardiogenic shock, typically in conjunction with revascularization. In rare cases, emergent heart transplantation may be required, particularly if CABG is not feasible or is unsuccessful and patients remain unstable despite maximal hemodynamic support.

Pharmacological therapy for P-SCAD is similar to standard SCAD therapy, as previously described. Intravenous heparin is routinely started for standard short-term myocardial infarction management but discontinued once SCAD is identified. Glycoprotein IIb/IIIa inhibitors and thrombolytics are contraindicated because of potential extension of intramural hematoma with these potent agents. Long-term medical therapy with aspirin and β-blocker (which reduces arterial wall stress) is appropriate. Optional agents include angiotensin-converting enzyme inhibitors (for left ventricular dysfunction), statins (for preexisting dyslipidemia), and nitrates (for heart failure, concomitant vasospasm, or residual coronary stenosis). Clopidogrel is mandatory for patients receiving coronary stents (for 1–12 months) and is commonly administered for a limited duration (<12 months) without PCI. There are no data for novel P2Y12 antagonists with SCAD. For patients treated conservatively, follow-up exercise testing should be considered, and repeat coronary angiography (or computed tomography if proximal/large arteries dissected) should be performed if ischemia is present to assess for subsequent revascularization necessity.

Natural History and Outcomes

Historical case reports and series suggested high mortality rates (38%–50%), which were likely influenced by limited options for intervention and early reporting bias of autopsy cases. Contemporary reports of patients surviving to cardiac catheterization with prompt diagnosis and management (including our 3 early postpartum cases) showed more favorable in-hospital outcomes. However, patients with P-SCAD appear to have more sinister presentations (both clinically and angiographically) compared with patients with non–P-SCAD. P-SCAD is associated with larger infarctions with higher peak troponin, worse left ventricular function, congestive heart failure, and cardiogenic shock. Recurrent coronary dissection in SCAD survivors is quite common, reported in 13% to 18% of larger series. It is not known if recurrent SCAD is higher in P-SCAD survivors. In a recently reported small series of SCAD survivors who subsequently became pregnant, 1 of 7 suffered recurrent SCAD at 9 weeks postpartum with LM dissection requiring CABG. Although this series was small, it highlights that recurrent SCAD can occur with subsequent pregnancies, and future pregnancies should be avoided in SCAD survivors given the potential severe complications.

Conclusions

P-SCAD is a rare but potentially catastrophic condition. We described 3 representative cases managed differently in the hospital (conservative treatment, PCI, and CABG) to demonstrate nuances of management decisions based on clinical presentation and coronary anatomy. Clinicians need to be familiar with angiographic appearances of SCAD for prompt diagnosis and with management strategies to appropriately risk stratify, treat, and follow up these patients.

Disclosures

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