

# Skin as a Window to Perioperative Risk in Systemic Sclerosis: Dermatologic Manifestations and Surgical Implications

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## ABSTRACT

### Purpose of Review

Systemic sclerosis (SSc) is a complex autoimmune connective tissue disease characterized by microvascular dysfunction, immune activation, and progressive fibrosis affecting the skin and multiple internal organs. As survival improves, an increasing number of patients with SSc are exposed to surgical procedures. This review aims to examine key dermatologic manifestations of SSc and explore their relevance to perioperative and postoperative risk, highlighting the role of the skin as a clinically accessible marker of surgical vulnerability.

### Recent Findings

Cutaneous involvement is the earliest and most consistent feature of SSc and reflects the severity of underlying vasculopathy and fibrosis. Manifestations such as skin thickening, Raynaud's phenomenon, digital ulcers, telangiectasias, calcinosis, and orofacial fibrosis are closely associated with cardiopulmonary disease, impaired wound healing, difficult airway management, and increased risk of perioperative complications. Emerging evidence supports the prognostic value of these dermatologic features as indicators of perioperative cardiovascular, pulmonary, infectious, and thrombotic risk.

### Summary

Dermatologic manifestations in systemic sclerosis extend beyond diagnostic relevance and provide valuable insight into perioperative risk. Integrating focused skin assessment into perioperative evaluation may improve risk stratification, guide anesthetic and surgical planning, and enhance postoperative surveillance. A structured, phenotype-based approach that incorporates dermatologic findings into multidisciplinary perioperative management may contribute to improved surgical outcomes in patients with systemic sclerosis.

### Keywords (MeSH terms)

Systemic sclerosis, Scleroderma, Cutaneous manifestations, Perioperative care, Anesthesia, Surgical risk, Pulmonary hypertension, Interstitial lung disease, Wound healing.

### Introduction

Systemic sclerosis (SSc) is a chronic autoimmune connective

tissue disease characterized by a complex interplay of small-vessel vasculopathy, endothelial dysfunction, immune activation, and progressive fibrosis affecting the skin and multiple internal organs, including the lungs, heart, kidneys, and gastrointestinal tract [1]. Although SSc is a rare disease, its clinical impact is substantial due to the high morbidity and mortality associated with cardiopulmonary involvement and the complexity of its

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multidisciplinary management [2].

The skin is the earliest and most consistently involved organ in systemic sclerosis and plays a central role in both disease classification and clinical assessment. Cutaneous thickening, microvascular abnormalities, telangiectasias, digital ulcers, calcinosis, and orofacial involvement not only reflect local fibrotic processes but also mirror the extent of underlying systemic vasculopathy and organ involvement [3]. In this context, the cutaneous phenotype may be regarded as a clinical “window” into disease severity and perioperative risk.

Despite this close pathophysiological relationship, the clinical approach to systemic sclerosis remains fragmented. Dermatologic manifestations are frequently assessed in isolation, primarily from a diagnostic or rheumatologic perspective, while perioperative risk is evaluated independently within anesthetic and surgical settings. This artificial separation has limited the recognition of the prognostic value of cutaneous findings as markers of perioperative risk, particularly with respect to cardiovascular, pulmonary, infectious, and technical complications during surgery [4].

Advances in the medical management of systemic sclerosis have led to improved survival, resulting in a growing number of patients undergoing elective and urgent surgical procedures [5]. In this evolving landscape, integrating dermatologic assessment into perioperative risk stratification is increasingly important, as many anesthetic and surgical challenges are foreshadowed by readily identifiable cutaneous features.

The aim of this review is to analyze the key dermatologic manifestations of systemic sclerosis and examine their relationship with perioperative and postoperative risk, proposing an integrated framework to optimize preoperative evaluation, anesthetic planning, and postoperative care in this complex patient population.

### **Shared Pathophysiology: From Skin Involvement to Perioperative Risk**

Systemic sclerosis is driven by a shared pathophysiological triad consisting of microvascular injury, immune-mediated inflammation, and progressive fibrosis. These processes develop in parallel within the skin and internal organs, explaining why cutaneous manifestations often precede or accompany cardiopulmonary and visceral involvement [6]. The skin therefore represents not only a target organ but also a visible marker of systemic disease activity.

Microvascular dysfunction is considered an early and central event in the pathogenesis of systemic sclerosis. Endothelial cell injury leads to impaired vasodilation, increased endothelin-1 production, reduced nitric oxide bioavailability, and chronic tissue hypoperfusion [7]. Recurrent ischemia–reperfusion injury promotes oxidative stress, platelet activation, and a prothrombotic state, ultimately resulting in structural vascular remodeling. These mechanisms underlie both cutaneous manifestations such as Raynaud’s phenomenon, digital ulcers, and telangiectasias, as

well as pulmonary arterial hypertension and myocardial ischemia, which are major determinants of perioperative risk [8].

Fibrosis represents the final common pathway of systemic sclerosis and affects the skin, lungs, heart, and gastrointestinal tract. Aberrant activation of fibroblasts and myofibroblasts, driven by profibrotic cytokines such as transforming growth factor- $\beta$  (TGF- $\beta$ ), platelet-derived growth factor, and interleukin-6, leads to excessive extracellular matrix deposition and tissue stiffening [9]. In the skin, this process manifests as progressive thickening and loss of elasticity, while in the lungs and heart it contributes to restrictive lung disease, impaired gas exchange, diastolic dysfunction, and arrhythmogenic myocardial fibrosis [10]. These fibrotic changes significantly limit physiological reserve and increase vulnerability to perioperative stress.

Endothelial dysfunction and fibrosis are further compounded by autonomic nervous system involvement, which is common in systemic sclerosis. Autonomic dysregulation contributes to impaired cardiovascular responses, orthostatic hypotension, abnormal heart rate variability, and altered thermoregulation [11]. During anesthesia and surgery, this impaired autonomic adaptability may predispose patients to hemodynamic instability, exaggerated responses to anesthetic agents, and difficulty maintaining adequate tissue perfusion.

Surgical stress acts as a potent amplifier of these underlying pathophysiological processes. The inflammatory and neurohumoral response to surgery can exacerbate endothelial injury, increase pulmonary vascular resistance, and worsen right ventricular function, particularly in patients with subclinical pulmonary hypertension or interstitial lung disease [12]. Additionally, perioperative hypoxia, hypothermia, and vasoconstriction may aggravate Raynaud’s phenomenon and compromise peripheral and wound perfusion, increasing the risk of ischemic and infectious complications.

Taken together, the same mechanisms responsible for cutaneous involvement in systemic sclerosis—vasculopathy, fibrosis, and endothelial dysfunction—also underpin the major cardiopulmonary and vascular complications encountered in the perioperative setting. Recognizing this shared pathophysiology provides a conceptual framework for understanding why dermatologic manifestations can serve as clinically meaningful indicators of perioperative risk.

### **Key Dermatologic Manifestations of Systemic Sclerosis and Their Clinical Significance**

Cutaneous involvement in systemic sclerosis is heterogeneous and evolves over time, reflecting the dynamic interplay between vasculopathy, inflammation, and fibrosis. Several dermatologic manifestations are particularly relevant in the perioperative setting, as they serve as visible markers of systemic disease severity and predict specific anesthetic and surgical challenges [13].

#### **Skin Thickening and Fibrosis**

Progressive skin thickening is the hallmark cutaneous

manifestation of systemic sclerosis and represents the most direct clinical expression of tissue fibrosis. Its extent and severity are commonly quantified using the modified Rodnan skin score (mRSS), which assesses skin thickness across 17 anatomical sites (Figure 1). Higher mRSS values have been consistently associated with diffuse cutaneous disease, increased risk of interstitial lung disease, cardiac involvement, and reduced survival [14].

From a perioperative perspective, extensive skin fibrosis is associated with reduced tissue compliance, impaired wound healing, and technical difficulties during patient positioning, vascular access, and regional anesthesia. Moreover, diffuse cutaneous involvement often parallels more aggressive systemic disease, signaling diminished cardiopulmonary reserve and increased vulnerability to perioperative stress [15].

### Raynaud’s Phenomenon and Digital Ulcers

Raynaud’s phenomenon is present in the vast majority of patients with systemic sclerosis and is frequently the earliest clinical manifestation. Chronic and severe vasospasm, compounded by structural vascular remodeling, predisposes patients to ischemic digital ulcers and pitting scars (Figure 2). Digital ulcers occur in up to 50% of patients and are strongly associated with diffuse disease, pulmonary arterial hypertension, and increased mortality [16].

In the perioperative setting, Raynaud’s phenomenon and digital ulcers indicate advanced microvascular disease and a heightened risk of ischemic complications. Perioperative hypothermia, vasoconstrictive agents, and hypotension may exacerbate digital ischemia, while the presence of active ulcers increases the risk of infection and impaired postoperative recovery [17].

### Telangiectasias

Telangiectasias represent a macroscopic manifestation of systemic microangiopathy and are commonly observed on the face, hands, lips, and mucosal surfaces (Figure 2). Their burden has been correlated with capillary loss, severe microvascular remodeling, and cardiopulmonary involvement, particularly pulmonary arterial hypertension [18].

Although often considered a cosmetic concern, extensive telangiectasias may signal advanced vascular disease and increased perioperative cardiovascular risk. In addition, mucocutaneous telangiectasias may predispose patients to bleeding complications during airway manipulation or invasive procedures [19].

### Calcinosis Cutis

Calcinosis cutis, characterized by the deposition of insoluble calcium salts in the skin and subcutaneous tissues, occurs predominantly in patients with long-standing disease and limited cutaneous systemic sclerosis (Figure 2). These deposits frequently affect pressure-prone areas such as the fingers, elbows, and forearms and may ulcerate or become secondarily infected [20].

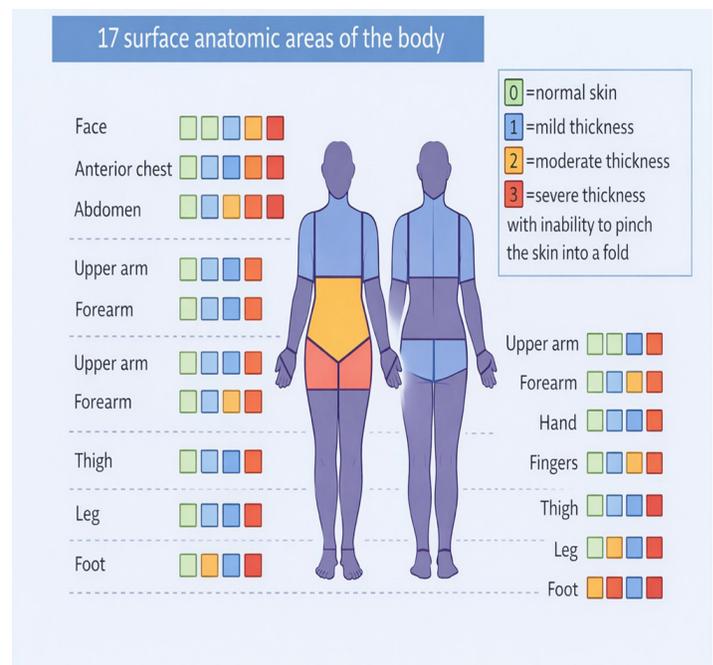
Calcinosis has important perioperative implications, including increased risk of local infection, impaired wound healing,

and technical challenges during surgical incisions or catheter placement. Chronic inflammation associated with calcinosis may further contribute to pain and delayed postoperative recovery [21].

### Orofacial Involvement and Microstomia

Fibrosis of perioral tissues leads to microstomia, reduced mouth opening, and loss of facial skin elasticity, often producing the characteristic “mask-like” facies of systemic sclerosis (Figure 2). Microstomia affects up to 80% of patients and may progress over time [22].

Orofacial involvement has direct anesthetic implications, as it is strongly associated with difficult mask ventilation and challenging airway management. Limited cervical mobility due to skin and soft tissue fibrosis may further compound these difficulties, increasing the need for advanced airway planning and specialized techniques [23].



**Figure 1:** Modified Rodnan Skin Score (mRSS) and cutaneous disease severity in systemic sclerosis.

Schematic representation of the 17 anatomical surface areas evaluated in the modified Rodnan skin score (mRSS). Each site is graded from 0 to 3 according to skin thickness, where higher scores indicate more severe cutaneous fibrosis. Increased mRSS values are associated with diffuse disease, greater systemic involvement, and higher perioperative risk.

Schematic representation of the 17 anatomical surface areas evaluated in the modified Rodnan skin score (mRSS). Each site is graded from 0 to 3 according to skin thickness, where higher scores indicate more severe cutaneous fibrosis. Increased mRSS values are associated with diffuse disease, greater systemic involvement, and higher perioperative risk.



**Figure 2:** Cutaneous manifestations of systemic sclerosis.

### Perioperative Implications of Dermatologic Manifestations in Systemic Sclerosis

The dermatologic manifestations of systemic sclerosis have direct and clinically relevant implications for perioperative management. Cutaneous findings frequently anticipate technical difficulties, anesthetic challenges, and postoperative complications, underscoring the importance of incorporating skin assessment into perioperative risk stratification (Figure 3).

#### Airway Management

Orofacial fibrosis, microstomia, reduced mandibular mobility, and cervical skin thickening are common in systemic sclerosis and represent major predictors of difficult airway management. Limited mouth opening and reduced cervical extension may complicate mask ventilation and direct laryngoscopy, increasing the likelihood of difficult or failed intubation. These challenges are often evident on preoperative physical examination, where perioral tightening and reduced interincisal distance reflect advanced cutaneous fibrosis (Figure 2).

In this context, a thorough preoperative airway assessment is essential, and anesthetic planning should include preparedness for advanced airway techniques, such as fiberoptic intubation or

videolaryngoscopy. Rapid sequence induction may be particularly hazardous due to the high prevalence of esophageal dysmotility and gastroesophageal reflux disease in patients with systemic sclerosis [24].

#### Vascular Access and Monitoring

Diffuse skin thickening and subcutaneous fibrosis may significantly impair peripheral venous access, complicating intravenous cannulation and arterial line placement. Cutaneous rigidity and loss of tissue elasticity may obscure anatomical landmarks and increase the risk of vascular injury [25]. These challenges are frequently underestimated unless specifically anticipated during preoperative assessment.

Early consideration of ultrasound-guided vascular access is recommended, particularly in patients with extensive skin involvement or prior access difficulties. Invasive hemodynamic monitoring may be required for high-risk procedures, especially in patients with suspected cardiopulmonary involvement, as outlined in the integrated perioperative algorithm (Figure 3).

#### Cardiovascular and Pulmonary Risk

Cutaneous markers of severe vasculopathy—such as extensive telangiectasias, digital ulcers, and high mRSS—are associated with an increased likelihood of pulmonary arterial hypertension, interstitial lung disease, and myocardial involvement. These conditions represent the principal drivers of perioperative morbidity and mortality in systemic sclerosis [26].

Patients with systemic sclerosis have an increased risk of perioperative myocardial ischemia, arrhythmias, and right ventricular dysfunction, particularly in the presence of pulmonary hypertension [27]. Similarly, interstitial lung disease reduces pulmonary reserve and increases susceptibility to hypoxemia, ventilator-induced lung injury, and postoperative respiratory failure. Recognition of these risks should prompt comprehensive preoperative cardiopulmonary evaluation and tailored intraoperative monitoring strategies (Figure 3).

#### Infectious and Thrombotic Risk

Chronic skin ulceration, calcinosis cutis, and impaired tissue perfusion predispose patients with systemic sclerosis to local and systemic infections. Digital ulcers and calcinosis-associated skin breakdown may serve as portals of entry for bacterial infection, increasing the risk of postoperative wound complications and sepsis.

In addition, systemic sclerosis is associated with a prothrombotic state, driven by endothelial dysfunction, chronic inflammation, and microvascular injury. Patients exhibit an increased risk of venous thromboembolism, particularly in the perioperative period [28]. Careful attention to thromboprophylaxis and early mobilization is therefore essential, especially in patients with active cutaneous vascular disease.

## Postoperative Considerations in Patients with Systemic Sclerosis

The postoperative period represents a particularly vulnerable phase for patients with systemic sclerosis, as surgical stress, immobilization, and residual anesthetic effects may exacerbate underlying vasculopathy, fibrosis, and cardiopulmonary dysfunction. Dermatologic manifestations observed preoperatively often anticipate specific postoperative complications and should inform surveillance and management strategies.

### Wound Healing and Skin Integrity

Impaired wound healing is a well-recognized complication in systemic sclerosis and is largely attributable to chronic microvascular dysfunction, reduced tissue perfusion, and fibrotic skin changes. Cutaneous thickening, digital ulcers, and calcinosis cutis are associated with delayed epithelialization, wound dehiscence, and increased susceptibility to surgical site infection [29].

Postoperative wound assessment should therefore be meticulous, particularly in areas of pre-existing skin involvement or pressure.

Strategies to minimize local ischemia—such as avoiding excessive tension on wound edges, maintaining normothermia, and optimizing oxygen delivery—are essential to reduce complications and promote healing.

### Pain Management

Pain control in systemic sclerosis is frequently challenging due to the coexistence of chronic pain syndromes, neuropathic pain, and ischemic discomfort related to vascular disease. Digital ulcers, calcinosis, and musculoskeletal involvement may contribute to baseline pain that is exacerbated in the postoperative setting [30].

Multimodal analgesia is recommended to minimize opioid exposure and reduce adverse effects, particularly in patients with cardiopulmonary compromise. Regional anesthesia techniques may be beneficial but should be carefully planned, as skin fibrosis and altered anatomy can complicate block placement. Additionally, careful monitoring for peripheral ischemia is warranted when vasoconstrictive agents are used for analgesia or hemodynamic support.

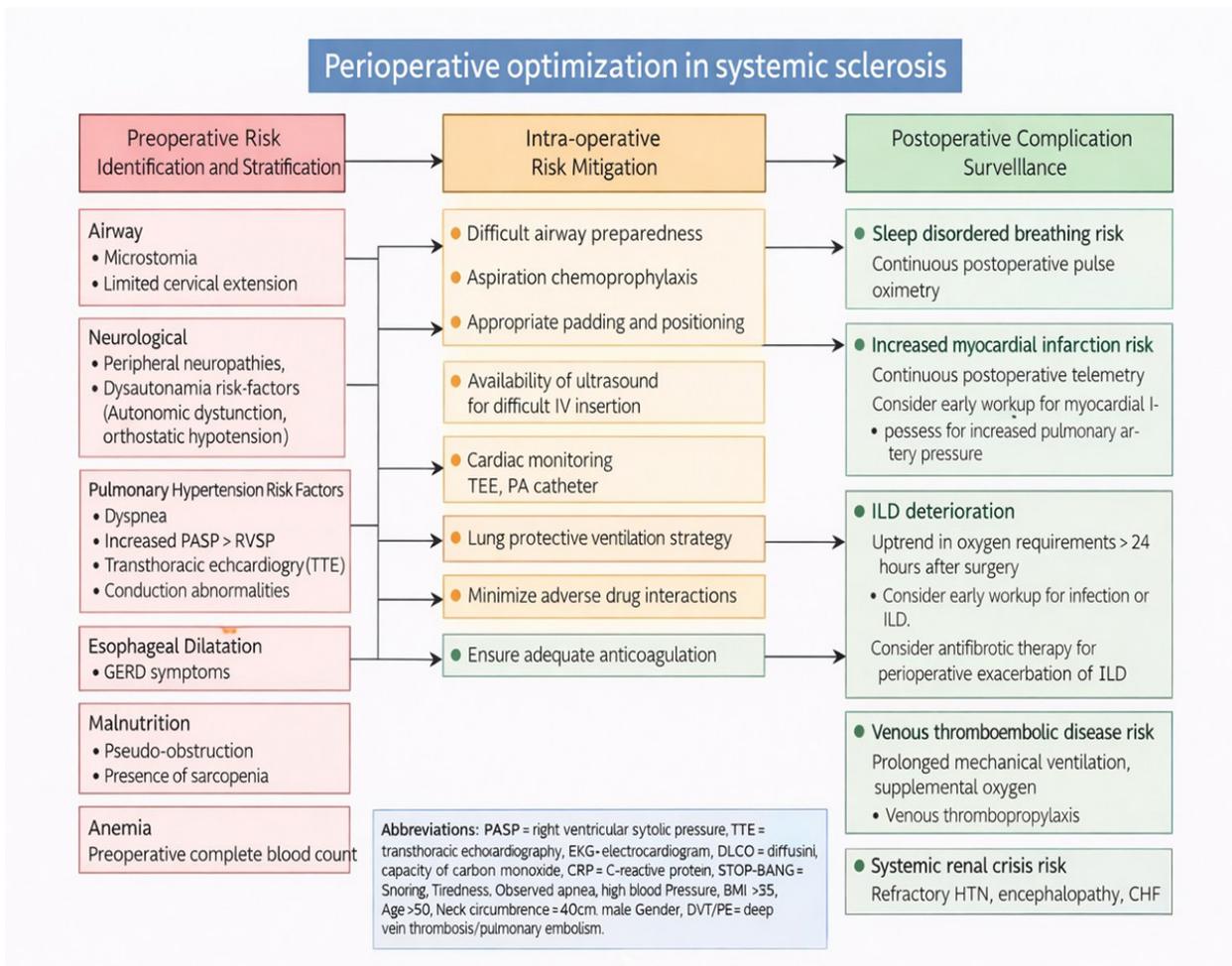


Figure 3: Perioperative optimization in patients with systemic sclerosis.

## Tissue Perfusion and Thermoregulation

Maintenance of adequate tissue perfusion is critical in the postoperative management of systemic sclerosis. Hypotension, hypovolemia, hypoxia, and hypothermia may all worsen microvascular dysfunction and precipitate ischemic complications, particularly in patients with severe Raynaud's phenomenon or active digital ulcers [31].

Postoperative hypothermia should be aggressively avoided, as cold exposure may trigger vasospasm and exacerbate digital ischemia. Active warming strategies and careful hemodynamic monitoring are especially important during the immediate postoperative period.

## Cardiopulmonary Surveillance

Cardiopulmonary complications are a leading cause of postoperative morbidity and mortality in systemic sclerosis. Patients with pulmonary arterial hypertension, interstitial lung disease, or myocardial involvement are at increased risk of postoperative hypoxemia, right ventricular failure, arrhythmias, and myocardial ischemia [32].

Continuous pulse oximetry, telemetry, and early mobilization should be considered in high-risk patients. A low threshold for postoperative imaging or cardiopulmonary reassessment is warranted in the presence of unexplained dyspnea, hypoxemia, chest pain, or hemodynamic instability. These considerations are summarized within the integrated perioperative management framework (Figure 3).

Schematic overview of perioperative management in systemic sclerosis, illustrating key components of preoperative risk identification and stratification, intraoperative risk mitigation, and postoperative complication surveillance. The algorithm integrates airway, vascular, cardiopulmonary, gastrointestinal, nutritional, and thrombotic risk factors to guide perioperative planning and postoperative monitoring in this high-risk population.

## Practical Proposal: Integrating Dermatologic Assessment into Perioperative Risk Stratification

Given the close pathophysiological relationship between cutaneous involvement and systemic disease severity in systemic sclerosis, a structured dermatologic assessment should be incorporated into routine perioperative evaluation. Such an approach allows early identification of high-risk patients and facilitates tailored anesthetic and surgical planning (Figure 3).

## Dermatologic Preoperative Checklist

A focused dermatologic checklist may serve as a practical and time-efficient tool during preoperative assessment. Key elements include:

- Extent and severity of skin thickening, ideally quantified using the modified Rodnan skin score (mRSS), with particular attention to diffuse cutaneous involvement.
- Presence of Raynaud's phenomenon, history of ischemic episodes, and active or prior digital ulcers.
- Distribution and burden of telangiectasias, particularly involving the face, lips, or mucosal surfaces.
- Evidence of calcinosis cutis, especially in areas relevant to surgical access or positioning.
- Orofacial involvement, including reduced mouth opening, perioral fibrosis, and limited cervical mobility.

These findings should not be viewed in isolation but rather as indicators of underlying vasculopathy, fibrosis, and potential cardiopulmonary involvement, prompting further targeted evaluation when appropriate [33].

## Risk-Based Perioperative Algorithm

Integration of dermatologic findings into a broader perioperative algorithm enables stratification of patients into low-, intermediate-, and high-risk categories (Figure 3). Patients with extensive skin fibrosis, active digital ulcers, marked telangiectasias, or microstomia should be considered at increased risk for:

- Difficult airway management,
- Impaired vascular access,
- Cardiopulmonary complications,

**Table 1:** Dermatologic findings in systemic sclerosis and their perioperative implications.

<i>Dermatologic finding</i>	<i>Pathophysiologic correlate</i>	<i>Perioperative implications</i>	<i>Recommended perioperative actions</i>
Extensive skin thickening (high mRSS)	Diffuse fibrosis, reduced tissue compliance, systemic disease severity	Difficult positioning, impaired wound healing, higher cardiopulmonary risk	Careful positioning, anticipate difficult access, enhanced monitoring
Raynaud's phenomenon	Severe microvascular dysfunction, vasospasm	Risk of perioperative ischemia, digital hypoperfusion, delayed healing	Avoid hypothermia, minimize vasoconstrictors, active warming
Active or prior digital ulcers	Advanced vasculopathy, chronic ischemia	Infection risk, poor wound healing, thrombotic risk	Inspect skin preoperatively, infection control, thromboprophylaxis
Telangiectasias (extensive or mucosal)	Microangiopathy, association with PAH	Bleeding risk, marker of cardiopulmonary involvement	Gentle airway manipulation, cardiopulmonary evaluation
Calcinosis cutis	Chronic inflammation, tissue hypoxia	Local infection, surgical access difficulty	Avoid incision through affected areas, meticulous wound care
Microstomia / orofacial fibrosis	Perioral and cervical fibrosis	Difficult mask ventilation and intubation	Advanced airway planning, consider fiberoptic techniques
Diffuse nailfold and digital changes	Peripheral vascular disease	Difficult IV access, monitoring challenges	Ultrasound-guided vascular access

- Delayed wound healing and infection.

In such cases, multidisciplinary planning involving anesthesiology, surgery, rheumatology, and, when indicated, cardiology or pulmonology is recommended. Early optimization, advanced airway planning, ultrasound-guided vascular access, and enhanced postoperative monitoring may mitigate perioperative morbidity [34].

By explicitly incorporating dermatologic assessment into perioperative decision-making, clinicians can move beyond a purely organ-based approach and adopt a more integrated, phenotype-driven strategy that reflects the systemic nature of the disease.

A focused dermatologic preoperative checklist, combined with an understanding of the perioperative implications of specific cutaneous findings, may improve risk stratification and perioperative planning (Table 1, Figure 3).

### Conclusions

Systemic sclerosis is a complex multisystem disease in which cutaneous manifestations reflect the underlying burden of vasculopathy, fibrosis, and endothelial dysfunction. Far from being isolated dermatologic findings, features such as skin thickening, Raynaud's phenomenon, digital ulcers, telangiectasias, calcinosis, and microstomia provide valuable insight into disease severity and perioperative risk.

This review highlights the concept of the skin as a clinically accessible "window" into perioperative vulnerability in systemic sclerosis. Dermatologic phenotypes anticipate many of the anesthetic and surgical challenges encountered in these patients, including difficult airway management, impaired vascular access, cardiopulmonary complications, and delayed wound healing.

Integrating dermatologic assessment into perioperative evaluation represents a practical and underutilized opportunity to improve risk stratification and optimize outcomes. A structured, phenotype-based approach—supported by multidisciplinary collaboration and guided by an integrated perioperative algorithm (Figure 3)—may enhance patient safety and reduce perioperative morbidity in this growing surgical population.

Future studies are needed to validate dermatology-informed perioperative risk models and to define standardized pathways that incorporate cutaneous assessment into routine surgical care for patients with systemic sclerosis.

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