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PREOPERATIVE EVALUATION (BJ SWEITZER, SECTION EDITOR)

An Update on Systemic Sclerosis and its Perioperative Management

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Abstract

Purpose of Review Systemic sclerosis or scleroderma (SSc) is a systemic, immune-mediated disease characterized by abnormal cutaneous and organ-based fibrosis that results in progressive end-organ dysfunction and decreased survival. SSc results in significant challenges for the practicing anesthesiologist due to its rarity, multi-system involvement, and limited evidence-based guidance for optimal perioperative care. In this update, we briefly discuss the recent evidence on the pathophysiology and current management of SSc, review the anesthesia-related literature, and extrapolate these observations into an optimal perioperative strategy for the care of SSc patients.

Recent Findings Evidence shows that patients with SSc demonstrate an increased risk for perioperative myocardial infarction, high rates of interstitial lung disease, pulmonary arterial hypertension, neurological disease, gastric dysmotility disorders, and challenging airway management, all findings that may result in suboptimal perioperative outcomes.

Summary Advances in SSc medical management have resulted in improved survival, likely increasing the number of patients who will be exposed to perioperative care. Optimal perioperative management and risk stratification should expand beyond the well-described airway challenges and consider numerous systemic manifestations of systemic sclerosis such as pulmonary arterial hypertension, interstitial lung disease, and cardiac sequelae.

Keywords Systemic sclerosis · Scleroderma · Anesthesia · Perioperative

Introduction

Systemic sclerosis is an autoimmune disorder characterized by small-vessel vasculopathy, abnormal collagen deposition with fibrosis, and autoantibody-mediated immune dysfunction characteristically observed in the integumentary system. In addition, progressive fibrosis of visceral organs such as the heart, the lungs, the kidneys, and the gastrointestinal tract results in substantial morbidity and mortality. Proposed mechanisms highlight the importance of small-vessel vasculopathy, with broad interplay of genetic and environmental triggers, as a causative factor for both cutaneous and visceral manifestations of SSc [1]. Ultimately, as small-vessel vasculopathy leads to recurrent ischemia-reperfusion injury, vessel elasticity is lost, and the pathognomonic development of fibrosis ensues, resulting in the classic features of SSc. First classified in 1988, it can be broadly divided into limited or diffuse cutaneous disease although many subtypes exist (limited cutaneous, diffuse cutaneous, sine, and overlap syndromes). The limited disease is of less severity than the diffuse, with less internal organ involvement and confinement of skin thickening to extensor surfaces of the body. Patients diagnosed with diffuse cutaneous SSc comprise 40% of patients and typically have proximal limb involvement, high rates of fibrotic lung changes, and increased risk for renal and cardiac involvement. Since 1980, SSc has been classified utilizing the American Rheumatological College criteria, followed by the LeRoy-Medsger criteria, and has undergone continuous refinement since that time [2, 3]. The latest iteration was refined in 2013

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in collaboration with the American College of Rheumatology and the European League against Rheumatism (ACR/EULAR). It demonstrates a 91% sensitivity and 92% specificity, and is summarized in Table 1 [4]. SSc is rare and demonstrates wide geographical variation. In the USA, the incidence is approximately 13.9/1,000,000 with a female to male ratio of 9.7:1 [5]. With improvements in management, survival in patients with SSc has improved over time, and the 10-year survival is 66% after diagnosis [6]. The EULAR Scleroderma Trials and Research database has demonstrated that 55% of deaths can be directly attributable to SSc and 41% are not directly associated with the disease [7]. Of the SSc-related deaths, visceral organ involvement account for most deaths, with pulmonary fibrosis (19%), pulmonary arterial hypertension (14%), and myocardial disease (14%) comprising the majority of causes. Of the non-SSc-related deaths, cancer (13%), infection (13%) and cardiovascular disease (12%) comprised most of the deaths.

Preoperative Considerations

Patients with SSc presenting for surgery can pose multiple challenges for the anesthesiologist, an observation that has been acknowledged in the early anesthesia literature [8]. Due to the rare incidence of SSc, few clinical studies have been published on perioperative care; therefore, preoperative evaluation should quantify the extent and severity of SSc-related disease manifestations and their perioperative implications. Fortunately, significant progress has been made in characterizing the general medical care of patients with SSc and this can be extrapolated to optimal perioperative management.

The frequency of neurological involvement is between 1 and 40% in SSc patients and can be categorized into distinct central, peripheral, and autonomic nervous system involvement. Patients with anti-Scl70 and U1RNP antibodies appear to have higher risk for neurological involvement [1, 9]. Epilepsy (42.6%) and headache (18.8%) predominate the central nervous system conditions observed in an SSc population [10]. In addition, a high psychiatric burden is also observed, particularly depression (30–40%) and anxiety (25–64%). Peripheral neuropathy related to nerve entrapment is common in SSc with peripheral sensorimotor polyneuropathy (14.25%), trigeminal neuropathy (16.52%), and carpal tunnel syndrome (6.56%) comprising the majority of diagnoses. Secondary Raynaud's phenomenon is a typical neurovascular complication in SSc and is characterized by collagen infiltration of distal blood vessels in the hand, rather than the classically described transient spasm of distal arteries. Severe cases may result in pain, numbness, ulcers, and loss of fingertips. At least one report describes severe pain with intravenous propofol administration in a patient with secondary Raynaud's phenomenon although this presumed association was controversial [11, 12]. Autonomic nervous system

Table 1 American College of Rheumatology/European League Against Rheumatism SSc classification criteria for the diagnosis of systemic sclerosis. Note the critical role that physical examination findings play in the diagnosis of SSc and the prominence of pulmonary arterial hypertension and interstitial lung disease

American College of Rheumatology/European League Against Rheumatism classification criteria for the diagnosis of systemic sclerosis†		
Systemic manifestations	Features	Score‡
Skin thickening of bilateral distal fingers extending to the metacarpophalangeal joints (sufficient criterion)	n/a	9
Skin thickening of the fingers	Puffiness	2
	Sclerodactyly evident between the MCP and PIP joints	4
Lesions on the tip of fingers (count the highest score)	Distal finger ulcers	2
	Distal finger pitting scar	3
Evidence of telangiectasia		2
Evidence of abnormal nailfold capillaries		2
Evidence of pulmonary arterial hypertension and/or interstitial lung disease (maximum score: 2)	Pulmonary arterial hypertension	2
	Interstitial lung disease	2
Evidence of Raynaud's phenomenon		3
Evidence of SSc-related autoantibodies (maximum score: 3)	Anti-centromere (3)	3
	Anti-topoisomerase I (3)	
	Anti-RNA polymerase III (3)	

MCP metacarpophalangeal joints, *PIP* proximal interphalangeal joints, *SSc* systemic sclerosis

Adapted from: van den Hoogen F, Kanna D, Fransen J et al. *Classification Criteria for Systemic Sclerosis: An American College of Rheumatology/European League Against Rheumatism Collaborative Initiative*. (2013)

‡Total score is determined by adding the maximum score achieved in each category. ≥ 9 is classified as definite SSc

disorders are common in SSc, and studies have reported an incidence of 79% for parasympathetic dysfunction and 55% for sympathetic dysfunction [13, 14]. SSc patients demonstrate a high incidence of parasympathetic autonomic instability as shown by orthostatic hypotension, blunted rise in blood pressure with sustained hand grip, and decreased compensatory heart rate variation with Valsalva maneuvers [15]. Neurological conditions should be carefully reviewed in all preoperative patients with SSc, with an emphasis on eliciting symptoms of autonomic dysfunction (e.g., orthostatic hypotension) and consideration of potential intraoperative positioning challenges for the chosen surgical procedure.

Cardiac involvement is prevalent in SSc. Brought upon by the sequelae of pathological extracellular matrix deposition, patients demonstrate a wide variety of cardiac conditions including progressive congestive heart failure, diastolic dysfunction, pericardial involvement, or arrhythmias. It may be classified as primary or secondary cardiac involvement. Secondary involvement is defined as the cumulative effect of age-related comorbid conditions in conjunction with progressive SSc-related fibrosis. SSc with cardiac involvement, particularly in those with the diffuse subtype of SSc, is related to higher mortality risk [16, 17••]. A three-fold increase in myocardial infarction has been documented in a retrospective matched incident cohort study of 1239 patients with SSc [18]. There is some evidence that SSc-related cardiac morbidity carries over to the perioperative period. For example, SSc-related sinoatrial involvement may result in perioperative dysrhythmias that may be enhanced by volatile anesthetics and other administered anesthetic drugs. This observation is highlighted by published reports of intraoperative cardiovascular events related to dysrhythmia and pulseless electrical activity [19–21]. Furthermore, one study has examined the association between SSc and perioperative cardiovascular risk for inpatient surgical procedures and found an independent association between SSc and perioperative myocardial infarction but not for ischemic stroke, cardiac arrest, or acute heart failure [22••]. Careful evaluation of the cardiovascular system, particularly in patients with risk factors for the development of SSc-related cardiomyopathy (male sex, diffuse SSc, presence of anti-topoisomerase and other antibodies, evidence of interstitial lung disease (ILD), and higher scores on the disability index) should be encouraged. An elicited history of dyspnea on exertion, palpitations, chest pain, or syncope should encourage the perioperative physician to pursue further characterization of SSc-related cardiac disease. Given the high incidence of congestive heart failure, dysrhythmias and pericardial disease, preoperative echocardiography, and electrocardiography (ECG) should be considered prior to moderate to high-risk surgical procedures.

SSc-related interstitial lung disease (ILD) and pulmonary arterial hypertension (PAH) are the two most common causes of SSc-related mortality and are believed to be related to vasculopathy of the small pulmonary arteries. PAH is rare with less than 1000 cases per year but occurs in approximately 10% of patients with SSc, commonly manifests with exertional dyspnea and results in a median survival of 3 years [23, 24]. Initial treatment usually involves sildenafil (class B evidence) to improve exercise tolerance and anticoagulation with warfarin (class B evidence). Newer treatments include endothelin antagonists and prostacyclin analogues. Little is known about the specific perioperative implications of SSc-related PAH although the anesthesiologist should be prepared to manage right ventricular dysfunction secondary to elevated pulmonary vascular resistance. Separately, pulmonary hypertension related to ILD or SSc-related cardiac involvement may increase both morbidity and mortality in the perioperative period and may be present in 8–12% of SSC patients [23, 25•]. ILD may present with mild changes in functional capacity before any radiographical evidence with the most common early change being a reduced forced vital capacity (FVC) with a normal forced expiratory volume, 1st second/forced vital capacity (FEV_1/FVC) ratio. Similar to pulmonary fibrosis due to other causes, the diffusing capacity for carbon monoxide (DLCO) correlates with the extent of gas exchange dysfunction [26]. Patients may present for surgery while being treated with disease-modifying agents such as cyclophosphamide, mycophenolate mofetil, and pirfenidone, and a high level of vigilance for potential drug interactions is warranted during the preoperative assessment. Select perioperative implications of routine disease-modifying agents in SSc have been summarized in Table 2.

Given the prominence of PAH and ILD in SSc-related mortality and morbidity, focused pulmonary assessment is useful for perioperative risk stratification and allows for early detection of perioperative complications. Reduced functional capacity in patients with scleroderma-related PAH and ILD may be detected through abnormalities in the 6-min walk test and the University of California in San Diego Shortness of Breath Questionnaire (UCSD SOBQ). The UCSD SOBQ has demonstrated responsiveness to negative changes in patients with both ILD and PAH [38]. The 6-min walk test remains the most commonly used tool to monitor functional capacity in patients with ILD. Its use has not been fully validated in this population, but it has demonstrated efficacy in the differentiation of clinically meaningful PAH and ILD in at least one study, lending its utility in preoperative risk stratification [39–41]. Radiographic pulmonary assessment should be considered in preoperative patients with symptoms of shortness of breath, dyspnea on exertion, or supplemental oxygen use. On high-resolution computed tomography (HRCT), SSc-related ILD is usually characterized as non-specific interstitial pneumonitis. Impulse oscillometry can detect subtle small airway dysfunction, and small airway abnormalities are found in 25% of SSc patients [42]. The small airway dysfunction was more common in patients with limited cutaneous SSc compared with the more severe diffuse SSc and was not appreciated on

Table 2 Select perioperative implications of medications used in the treatment of systemic sclerosis. Substantial progress has been made in the treatment of SSc using disease-modifying agents [27–37]

Medication	SSc use	Perioperative implications
Calcium channel blockers	Raynaud's phenomenon	Generally beneficial; reduces cardiac morbidity after non-cardiac surgery[25]
Cyclophosphamide	ILD	Immunosuppression; drug-related cardiomyopathy
Mycophenolic acid	ILD	Immunosuppression, potential enhancement with PPI[26]
Azathioprine	ILD	Transient antagonism of atracurium, vecuronium, and pancuronium[27].
Epoprostenol	PAH	↑Risk of blood loss in LVAD placement[28]
Endothelin receptor antagonists*	PAH	May impair vasoconstriction during hemorrhage[29]. Negative lusitropic myocardial effects[30]
Phosphodiesterase-5 inhibitors**	PAH, digital ulcers	Risk of ischemic optic neuropathy[31], reduction in biventricular systolic function[32], cerebral venous thrombosis[33]
Glucocorticoids	Reduce SSc lesion inflammation	May precipitate scleroderma renal crisis
Methotrexate	Inflammatory arthritis	Thrombocytopenia; interstitial pneumonitis; ↑pneumonia; possible protective effect for MACE[34]
Chelating agents†	SSc calcinosis, fibrosis	Nephropathy, aplasia, polymyositis (penicillamine)
Bisphosphonates‡	SSc calcinosis	Modest ↑risk of atrial fibrillation[35]
Rituximab	Skin fibrosis	Infusion-related reactions; progressive multifocal leukoencephalopathy; mucocutaneous reactions

ILD interstitial lung disease, *PPI* proton pump inhibitor, *LVAD* left ventricular assist device, *PAH* pulmonary arterial hypertension, *SSc* systemic sclerosis, *MACE* major adverse cardiovascular events

*Ambrisentan, bosentan, macitentan

**Sildenafil, tadalafil, vardenafil, avanafil

†Penicillamine (less common)

‡Alendronate, ibandronate, risedronate, zoledronic (no longer recommended in SSc)

↑increased

standard pulmonary function tests. Large airway obstruction was rare, but common defects in pulmonary function testing, such as reduced DLCO and total lung capacity (TLC), correlate with worsening SSc lung involvement. This suggests that a high level of vigilance for the presence of reactive airway disease should be considered in patients with SSc even when asymptomatic and that preoperative pulmonary function testing may provide useful guidance for preoperative optimization. In addition, rising inflammatory markers such as an erythrocyte sedimentation rate of ≥ 20 mm/h and C-reactive protein ≥ 5 mg/L are correlated with deterioration in pulmonary function tests and pulmonary arterial hypertension, suggesting their utility in the preoperative assessment of patients with SSc [43].

Esophageal dysmotility disorders feature prominently in SSc with dysphagia and gastroesophageal reflux disease (GERD) occurring in 70% of patients [44]. Gastric involvement and hypotonic lower esophageal sphincter tone result in impaired emptying and food stasis. Due to the potential risk of perioperative aspiration, these abnormalities should be taken into consideration when selecting the anesthetic technique. Other severe gastrointestinal manifestations, including pseudo-obstruction, intestinal dysmotility disorders, volvulus, and intestinal telangiectasia, may signal higher mortality risk and potentially increase the perioperative risk profile [45, 46]. Gastrointestinal manifestations may also indicate the presence of malnutrition (20% of patients), sarcopenia, vitamin C deficiency, and worsening lung disease [47, 48]. SSc-related primary biliary cirrhosis (PBC) has been shown to occur in 2–22%, but is associated with improved outcomes compared with other forms of PBC [49, 50]. Regardless, the presence of PBC alone has not been shown to be associated with increased perioperative morbidity or mortality [51].

SSc-related renal impairment occurs in approximately 4–6% of patients, with the worst outcomes being encountered in patients with scleroderma renal crisis (SRC), a form of severe renal decompensation [52]. The adoption of angiotensin-converting enzyme inhibitors (ACEIs) has improved outcomes in SRC but overall survival remains poor, particularly if the disease is severe enough to necessitate dialysis [53]. Its triggers are unclear, but characteristic presentations of SRC include features of malignant hypertension, encephalopathy, and evidence of congestive heart failure.

SSc patients have an increased risk of unprovoked venous thromboembolic disease (VTE), likely related to the underlying inflammatory state and its pro-thrombotic nature [54–56]. Although the specific trigger for increased VTE risk is as of yet unclear, increased presence of antiphospholipid antibodies has been observed in this population, possibly contributing to the high rate of venous thrombosis [57]. The highest risk was found in patients within 12 months of SSc diagnosis and is

strongly associated with female sex, atrial fibrillation, and heart failure. Trauma and glucocorticoid use were independently associated with DVT, although surgery alone was not, which may be related to the common use of prophylactic anticoagulation in the perioperative period. Anemia, likely related to marrow aplasia or malnutrition, is common in patients with SSc [58]. In conclusion, SSc is a disease process with numerous perioperative implications of which the most clinically relevant findings are summarized in Fig. 1.

Intraoperative Considerations

Few studies offer guidance on the specific perioperative care of SSc patients and many of these are based on observations from case reports or series. Incidental findings of difficult intravenous access in case reports of systemic sclerosis are common [59–61]. Due to skin thickening, intravenous access may pose a challenge, and it is reasonable to have a low threshold for ultrasound guidance for vascular catheter

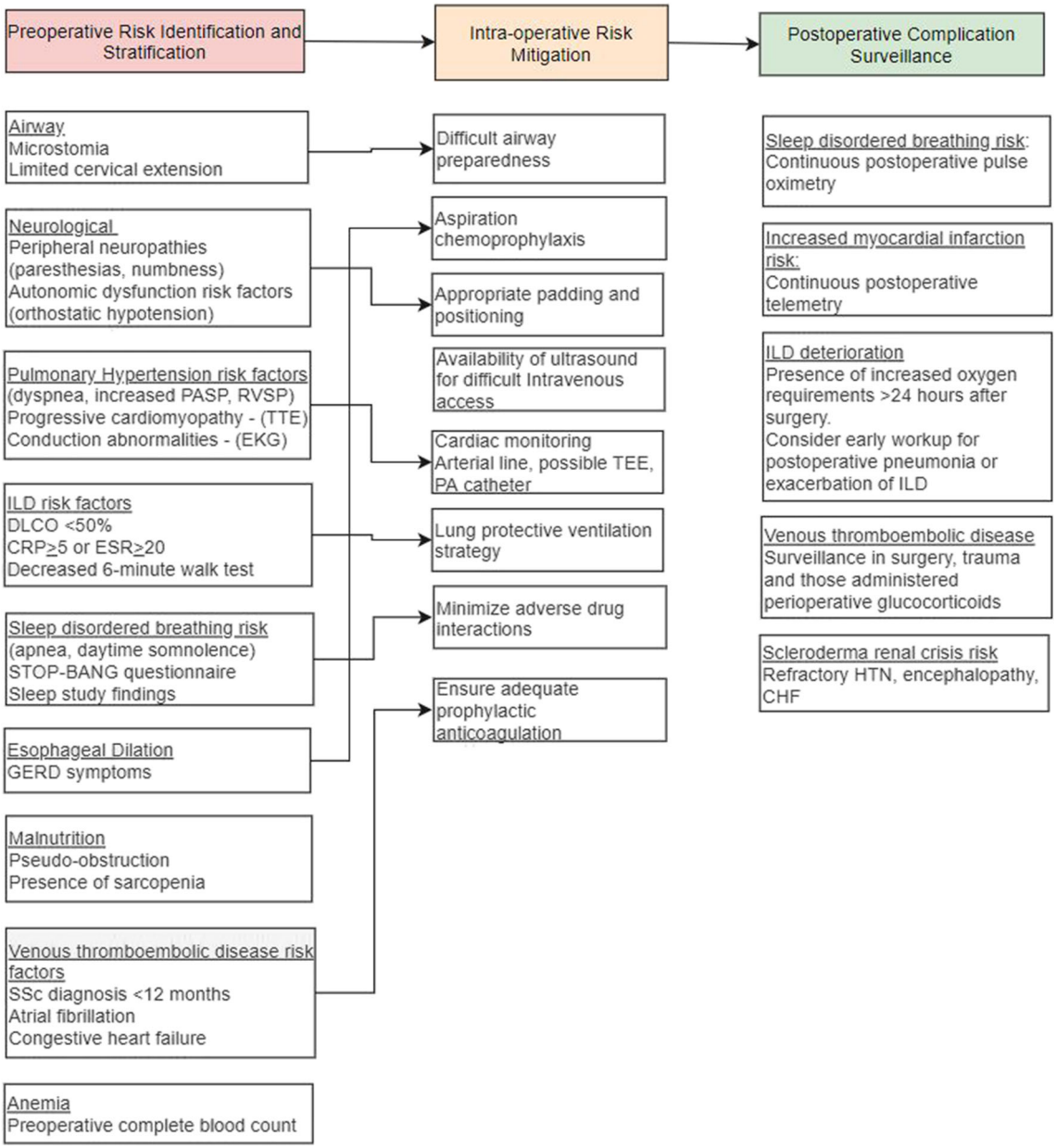

Fig. 1 Perioperative optimization in systemic sclerosis. The salient points in the perioperative optimization of patients with SSc are highlighted

insertion [62]. Airway management may pose a problem in SSc. There are reports of microstomia and restricted mouth opening, likely related to temporomandibular joint fibrosis, and fibrotic changes of the neck resulting in poor cervical extension [63, 64]. In addition, reports have emphasized the presence of limited cervical extension as an indicator of both difficult mask ventilation and need for fiberoptic bronchoscopy assisted intubation [63]. The recent data on airway management is summarized in Table 3. In addition, since approximately 90% of SSc patients have GERD and other gastrointestinal dysmotility disorders, a higher than normal aspiration risk is likely [65]. Indeed, a high prevalence of esophageal dilation is common and may be associated with an increased risk of centrilobular fibrosis of the lung, emphasizing the complex interplay between the pathophysiologic findings in SSc [66]. Appropriate preoperative precautions such as the administration of a histamine-2 (H_2) blocker and when appropriate, preoperative nasogastric suctioning, should be strongly considered. This recommendation is particularly strong since aspiration events are independently associated with risk of death in hospitalized SSc patients [67]. Nasogastric tube placement is performed with caution due to the high incidence of esophageal stricture disease in patients with SSc, increasing the already high-risk of esophageal perforation [68]. In addition, esophageal pathology may complicate placement of a transesophageal echocardiography (TEE) probe. Acknowledging the risk of difficult mask ventilation and endotracheal intubation, a rapid sequence intubation may not be advisable, and careful risk mitigation of aspiration should be considered before induction of general anesthesia. The potential perioperative risk for peripheral neuropathies should be mitigated with appropriate positioning and padding.

Given the common occurrence of cardiac involvement in SSc, intraoperative care should focus on close hemodynamic monitoring commensurate with the complexity of the surgical procedure. Dysrhythmias, likely drug enhanced, have been reported in patients with SSc and may lead to increased perioperative risk. In patients with pre-existing PAH or PH (pulmonary hypertension), poor right ventricular performance will likely be exacerbated by high intraoperative fluid loads, metabolic acidosis, and surgical stress resulting in deteriorating hemodynamics. Although little evidence is available to support their use, invasive monitoring may be advisable, dependent on the procedure and severity of disease. Thus, the benefits and risks should be carefully weighed before placement of pulmonary artery catheters and TEE probes in SSc patients. The severity of the PAH or PH may require inotropic agents (sympathomimetic and non-sympathomimetic vasopressors), vasodilators such as nitric oxide, and inhaled prostacyclin analogues.

Substantial ventilator-related harm may be induced in SSc patients with ILD, and efforts should be made to minimize the use of invasive mechanical ventilation. This observation is

Table 3 A limited synopsis of airway management in the reported literature since 1990. Microstomia is likely a prominent physical feature of challenging airway management in the patient with SSc

Study	*n*	Comment	Mallampati classification	Microstomia	Limited cervical extension	Difficult laryngoscopy	Other airway adjunct	Final airway instrument
Shionoya Y et al. (2020)	1	Avoided endotracheal intubation		y				n/a
Kanter GJ, Barash PG (1998)	1	Unanticipated difficult airway	1		y	y	y	Transtracheal wire guided
Bailey AR et al. (1999)	1	Successful spinal anesthesia	4	y				n/a
D'Angelo R, Miller R (1997)	1	Awake fiberoptic intubation		y	y		y	Fiberoptic bronchoscope
Moaveni JC et al. (2015)	2	Spinal anesthesia d/t favorable airway	3	y	n			n/a
		Awake fiberoptic intubation	4	y	y		y	Fiberoptic bronchoscope
Ye F et al. (2016)	1	Pre-emptive Shikani intubating stylet	3	y	n		y	Shikani intubating stylet
Bansal T, Hooda S (2013)	1	Refused awake fiberoptic intubation	3	y	y	n	n	Direct laryngoscopy

y yes; *n* no

supported by a meta-analysis of 15 studies that examined risk factors for intensive care unit–related mortality in a mixed ILD population and found a strong correlation between mortality and the application of invasive mechanical ventilation [69]. A study of SSc-related intensive care admission outcomes found higher mortality in patients with diffuse cutaneous SSc (72.5%), pulmonary fibrosis (71.7%), and/or pulmonary hypertension (31.5%) [70]. In-hospital mortality and 6-month mortality were 31.8% and 39.0%, respectively, and the majority were admitted with respiratory failure. This study also found a strong correlation between invasive mechanical ventilation and mortality. Another study found that a DLCO < 50% is a strong predictor for cardiopulmonary-related hospitalization in patients with SSc and pulmonary hypertension [71•]. Furthermore, acute exacerbations of ILD may also be more common in SSc compared with other ILD-related conditions and show increased mortality with invasive ventilation [72]. Although specific guidance concerning mechanical ventilation in SSc-related ILD is lacking, prudence dictates the use of perioperative lung protective ventilation strategies [73].

Given the evidence that invasive ventilation may contribute to SSc-related morbidity, it is reasonable to extrapolate that the appropriate use of regional or neuraxial techniques should be considered in the care of this patient population when possible. Regional and neuraxial techniques have been successfully performed in patients with SSc but may be technically challenging [74–76]. Furthermore, autonomic nervous system dysfunction may exacerbate hemodynamic effects of neuraxial anesthesia. Uneventful spinal anesthesia for cesarean section has been reported, but severe hypotension requiring treatment has been noted [60, 77, 78]. Prolonged sensory blockade has been reported and may be related to tissue pH difference reducing the unionized fraction of local anesthetic, or peripheral vascular insufficiency at the regional block location causing reduced clearance [75, 79, 80]. Furthermore, the increasing application of ultrasound guidance has likely reduced the need for high-volume local anesthetic for regional anesthesia, further reducing the risk of prolonged block. It is highly probable that obstetric anesthesia is safe, but parturients with SSc should be considered high-risk, necessitating a high level of preparedness and multi-disciplinary team involvement. Furthermore, the limited data suggests that, though it may be technically difficult, neuraxial and regional anesthesia are safe and should be encouraged in patients with SSc for all eligible procedures particularly with ultrasound guidance.

Postoperative Considerations

Postoperative vigilance remains the mainstay of safe perioperative care. Several aspects of postoperative monitoring should be considered after surgical intervention. SSc patients may be at risk for postoperative sleep-disordered breathing has been reported with 31.2% of patients having abnormal pulse oximetry readings on sleep studies [81]. This suggests that SSc patients may benefit from close postoperative ventilatory monitoring. Secondly, the increased SSc-specific risk for myocardial infarction should trigger close postoperative scrutiny. General principles for the care of SSc patients with PH or PAH continue through the postoperative time period. Meticulous attention to maintaining sinus rhythm, avoiding right ventricular stress, avoiding excessive hypercarbia, and early correction of metabolic abnormalities are important. Given the high incidence of ILD, symptoms of acute postoperative respiratory worsening, postoperative pneumonia, or acute exacerbation of ILD are addressed expeditiously. These adverse events are potentially high-mortality events in patients with SSc. The relationship of SRC and perioperative care is unclear, but close monitoring, particularly for signs and symptoms of refractory hypertension and aggressive management of deteriorating kidney function, should be considered. Furthermore, aggressive venous thromboembolic prophylaxis should be considered in most SSc patients, particularly those with a recent (< 12 months) diagnosis, atrial fibrillation, or cardiomyopathy.

Conclusion

Recent advances in the understanding and management of SSc have resulted in substantial improvements in survival. As many SSc patients continue to age with comorbidities such as PAH and ILD, it is likely that many will present for surgical interventions. Given its systemic involvement, the anesthesiologist needs to carefully weigh the implications of anesthetic techniques, and their potential for interactions with the underlying pathophysiology of SSc. Secondly, patients with systemic sclerosis often have well-compensated mechanisms for their underlying disease, which may be unmasked with anesthetic interventions. A difficult airway and a potential risk for aspiration should be anticipated, and appropriate preoperative preparation should be emphasized. Furthermore, SSc patients are at risk for postoperative myocardial infarction, and prolonged mechanical ventilation may result in substantial morbidity, specifically in those patients with ILD. Many questions remain, and further research is needed to better identify perioperative risk factors and determine optimal perioperative strategies in the care of these complex patients.

Availability of Data and Materials N/a

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Compliance with Ethical Standards

Conflicts of Interest None of the authors has any potential conflicts of interest to disclose.

Ethics Approval N/a

Human and Animal Rights and Informed Consent This article does not contain any studies performed by the co-authors that had human or animal subjects.

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