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Lung Transplantation in Systemic Sclerosis: a Practice Survey of United States Lung Transplant Centers

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Background. Lung transplantation in patients with systemic sclerosis (SSc) can be complicated by extrapulmonary manifestations of the disease, leading to concerns regarding posttransplant complications and outcomes. **Methods.** We conducted a web-based survey of adult lung transplant programs in the United States regarding their practices in patients with SSc. **Results.** Sixty percent (37/62) of the eligible centers responded to the survey, majority of the respondents were medical directors (81%). Most centers would consider transplanting patients with mild or moderate esophageal disease (92% or 75%, respectively) or gastroparesis (59%). A minority would consider patients with severe esophageal dysmotility (37%), digital ulcers (21%), or low body mass index (19%). Most centers conducted extensive pretransplant gastrointestinal evaluation and use a conservative feeding approach with prolonged nothing by mouth (83%) and postpyloric feeding (89%). Antireflux surgery is commonly considered (40%) with partial fundoplication being the procedure of choice (67%). Most respondents expected similar outcomes of acute or chronic rejection (81% and 51%, respectively), respiratory infections (76%), and 1-year survival (70%). **Conclusions.** Most US lung transplant centers do not universally exclude SSc from lung transplant listing, but most support extensive pretransplant gastrointestinal testing and a conservative approach to feeding in the early posttransplant period.

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Systemic sclerosis (SSc) is a systemic autoimmune disease that simultaneously affects multiple organs. Lung involvement occurs in about 80% of patients, predominantly presenting as interstitial lung disease (ILD) or pulmonary arterial hypertension (PAH).[1] Lung disease can cause severe morbidity and is the most common cause of death in SSc patients.[2] Depending on the stage and phenotype of the disease, treatment options include immunosuppressants, antifibrotic agents, pulmonary vasodilators, and autologous stem cell transplantation. Lung transplantation is a therapeutic option in progressive, end-stage lung disease.[3]

Lung transplantation in SSc patients is often considered high risk. Concerns regarding extrapulmonary disease and posttransplant complications may prevent some transplant centers from considering lung transplant in patients with SSc. Significant concerns include esophageal dysmotility, vascular disease, increased allosensitization, renal disease, and poor nutritional status.[4,5] Involvement of the gastrointestinal (GI) tract is seen in >90% of patients with SSc, often leading to oropharyngeal dysphagia, esophageal dysmotility, severe gastroesophageal reflux disease (GERD), and gastroparesis.[6,7] GERD and microaspiration are associated with impaired posttransplant survival and increased incidence of chronic lung allograft dysfunction.[8] Vascular disease leading to ischemic digital ulcers has been reported in 40% of SSc patients, this can lead to critical limb ischemia, necrosis, and infections in the posttransplant period.[5] Malnutrition in SSc patients is common and

is a risk factor for early postlung transplant mortality and reduced quality of life.[9,10]

Despite these concerns, several contemporary single and multicenter studies have demonstrated equivalent posttransplant outcomes in patients with SSc compared to other indications.[11-14] A study of US lung transplant recipients analyzing data from the United Network for Organ Sharing database, reported a higher 1-year mortality in SSc patients compared with non-SSc–ILD patients, but similar to non-SSc–related PAH.[13] The clinical practices in the published studies vary widely in the pretransplant evaluation as well as posttransplant care. While some programs complete extensive pretransplant GI testing, others only perform GI testing on symptomatic patients.[11-14] Given the risk for aspiration, some programs have a protocol of keeping patients nothing by mouth (NPO) with postpyloric feeding and early antireflux surgery, while other programs allow for early oral intake with aspiration precautions.[11-14]

Guidelines regarding workup and management of SSc patients prelung and postlung transplant are lacking. The International Society for Heart and Lung Transplantation consensus statement from 2015 recommends that carefully selected patients with SSc be considered for lung transplant.[15] A position paper from a multidisciplinary working group in France made more detailed recommendations regarding candidate selection and management of SSc patients.[5] However, SSc is considered a relative contraindication at several lung transplant programs in the United States and practices vary widely based on individual experiences and preferences. The objective of this survey is to determine the practices of US lung transplant programs in the evaluation and management of patients with SSc.

MATERIALS AND METHODS

The study was approved by the Temple University Institutional Review Board (Protocol number 26654) and adheres to the ethical standards and principles of the Declarations of Helsinki.

Study Participants

We surveyed physician representatives from all active, adult lung transplant centers in the United States. Lung transplant centers in the United States registered in the Scientific Registry of Transplant Recipients (SRTR) database that performed >1 lung transplant in 2018 were identified.[16] The medical and surgical directors and transplant pulmonologists were identified from the program website. The survey was conducted between May 15 and July 31, 2020. Lung transplant center medical directors were emailed an invitation with a unique link to the survey. The survey was first emailed to the medical director. If no response was obtained after 7 d, 2 additional attempts were made weekly. Alternate physician representatives that included the surgical director and transplant pulmonologists were contacted if no response was received by 4 wk. Following a similar protocol, a third individual was approached when possible. In instances of 2 or more responses, only one response from every program was recorded, prioritizing the medical director then surgical director then transplant pulmonologist. Data from SRTR about the number of adult lung transplants performed in 2018 were collected and were linked to the survey responses.[5]

Survey Instrument

The survey content was developed by the investigators based on literature review, experience, and stakeholder assessment. The survey questions were developed and refined through an iterative process for format and usability. The American College of Chest Physicians Transplant Network Steering Committee members gave critical feedback. The survey addressed individual transplant center's practices regarding the evaluation and management of patients with SSc (Online Supplement, **SDC**, http://links.lww.com/TXD/A362). The survey was divided into 4 sections: background information, pretransplant evaluation/candidacy, posttransplant management, and opinions about posttransplant outcomes. The survey was administered using the web-based platform Research Electronic Data Capture database.[17]

Survey questions were fact-based with multiple-choice responses. Responses to most questions included a Likert scale. Respondents could respond to as many or as few questions that they felt best able to contribute.

No incentive was provided for completing the survey. The survey responses were stored in Research Electronic Data Capture database hosted by Temple University.[17] The responses were linked to the center to track responses; however, they were deidentified for analysis.

Data Analysis

One survey per transplant center was analyzed. Completed surveys were analyzed preferentially over partially completed surveys. If >1 survey with the same degree of completion was received, the medical director's response was preferentially analyzed. Partially completed surveys and surveys with omitted questions were included in the analysis if no completed survey was received from the transplant center.

We used descriptive statistics to report findings. Results are presented as percentage of respondents to the particular question unless otherwise indicated. We compared responses between low-volume centers (< 20 transplants/y), medium-volume centers (20–39 transplants/y), and high-volume centers (> 40 transplants/y). The Fisher exact test was used to compare responses between the 3 groups. A P value of < 0.05 was considered as statistically significant. All statistical analyses were performed with the use of Stata 14.0 (StataCorp LP, College Station, TX).

RESULTS

Respondent and Program Characteristics

Of the US adult lung transplant centers, 60% (37/62) completed the survey. We did not receive multiple response from any center and thus did not have to preferentially analyze any responses. The majority of respondents were medical directors (81%) with transplant pulmonologists (13%) and surgical directors (5%) making up the rest of respondents. The median number of lung transplants performed by each center per SRTR data were 31 (interquartile range, 19–49) with a total of 1384 transplants in 2018 for all indications. All Organ Procurement and Transplantation Network regions except region 1 (New England) and region 9 (New York and Vermont) were represented. Respondents included representatives from low-, medium-, and high-volume centers. The number of patients with SSc that received a lung transplant at each center ranged from 0 to <10 during the year 2018 per respondent estimates: 40% reported no SSc transplants, 54%

between 1 and 5 SSc transplants, and 5% between 6 and 10 SSc transplants.

Consideration for Transplant Candidacy

The diagnosis of SSc was considered a contraindication for lung transplantation by 8% (3/37) of centers. The reasons for this consideration were concern about higher 1-y mortality (67%), prior experience with poor outcomes (67%), concern for GI disease leading to rejection (33%), and lack of supportive services (33%; Figure 1).

Regarding GI involvement with SSc, most centers were likely or extremely likely to consider candidates with mild esophageal disease (92%), moderate esophageal disease (76%), or gastroparesis (59%) for transplant listing. However, only 38% of centers were likely or extremely likely to consider patients with severe esophageal dysmotility. Regarding other SSc-related considerations, most centers were likely or extremely likely to transplant patients with an elevated panel reactive antibodies (65%) or severe PAH (78%), but fewer centers were likely or extremely likely to transplant patients with vascular disease or digital ulcers (22%) or low body mass index (BMI; 19%).

When comparing candidate selection practices between low-volume (<20 transplants/y), medium-volume (20–39 transplants/y), and high-volume (>40 transplants/y) centers, we did not find a difference in candidate selection practices between the 3 groups (Table 1).

Pretransplant Evaluation

Most centers conduct an extensive GI evaluation even in the absence of symptoms in patients with SSc. Centers were extremely likely or likely to conduct barium esophagogram (95%), esophageal manometry (92%), pH probe and impedance testing (92%), gastric emptying study (70.2%), and formal evaluation by a gastroenterologist (54%; (Figure 2).

Early Posttransplant Management

In patients with SSc and severe esophageal disease, most centers were extremely likely or likely to have a conservative feeding practice of prolonged NPO for greater than 2 wk (84%) and postpyloric feeding tube before hospital discharge (89%). Repeat GI testing was extremely likely or likely to be done by 59% of the centers (Figure 3).

Antireflux Surgery

A minority of centers (40%) reported they were extremely likely or likely to perform antireflux surgery in patients with severe esophageal disease. When asked about the surgical modality of choice, centers were extremely likely or likely to prefer partial fundoplication (65%). Only a small number of centers reported being likely to do a complete fundoplication (8%) and gastric bypass (11%). The timing of antireflux surgery varied widely. Most centers were extremely likely or likely to perform surgery 3 to 6 mo (50%) or beyond 6 mo (42%) posttransplant. The remaining centers were likely or extremely like to perform antireflux surgery before transplant (26%) or 0 to 3 mo (38%) posttransplant (Figure 4).

Posttransplant Outcomes

Respondents at centers that perform lung transplants on patients with SSc were asked about their impression regarding lung transplant outcomes of patients with SSc compared with other transplant indications. Most respondents believed the incidence of acute rejection (81%), respiratory infections (76%), and chronic rejection (51%) were similar to other indications. Regarding posttransplant survival, 70% reported similar 1-y survival and 49% reported similar 5-y survival. However, most respondents (54%) believed that patients with SSc have a worse quality of life compared to other indications (Figure 5).

DISCUSSION

Lung transplant for the SSc-related ILD or SSc-related PAH account for only 2% of all lung transplants in the United States.[18] It is unknown how much this low prevalence reflects decreased transplant opportunity according to selection criteria and, if so, which criteria are utilized for exclusion.

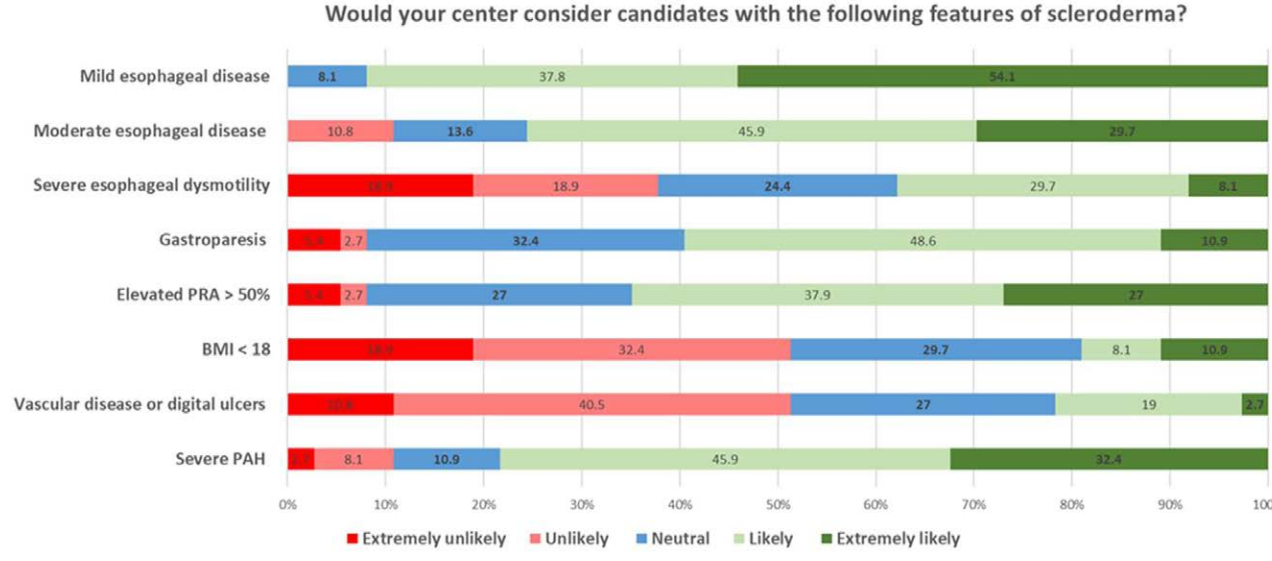

FIGURE 1. Consideration for transplant candidacy. BMI, body mass index; PAH, pulmonary arterial hypertension; PRA, panel reactive antibody.

TABLE 1.

Opinion regarding candidacy: number of centers with response of extremely likely/likely/neutral to the question 'would your center consider candidates with the following features of scleroderma?'

		Transplants per y			
	Total	Low volume 0–19 (n = 10)	Moderate volume 20–39 (n = 12)	Large volume > 40 (n = 15)	*P*
Mild esophageal disease, N (%)					
Extremely likely/likely/neutral	37 (100.0)	10 (100.0)	12 (100.0)	15 (100.0)	
Moderate esophageal disease, N (%)					0.66
Extremely likely/likely/neutral	33 (89.2)	8 (80.0)	11 (91.7)	14 (93.3)	
Unlikely/extremely unlikely	4 (10.8)	2 (20.0)	1 (8.3)	1 (6.7)	
Severe esophageal dysmotility, N (%)					0.69
Extremely likely/likely/neutral	23 (62.2)	5 (50.0)	8 (66.7)	10 (66.7)	
Unlikely/extremely unlikely	14 (37.8)	5 (50.0)	4 (33.3)	5 (33.3)	
Gastroparesis, N (%)					0.34
Extremely likely/likely/neutral	34 (91.9)	8 (80.0)	12 (100.0)	14 (93.3)	
Unlikely/extremely unlikely	3 (8.1)	2 (20.0)	0 (0.0)	1 (6.7)	
Elevated panel reactive antibodies > 50%, N (%)					0.17
Extremely likely/likely/neutral	34 (91.9)	8 (80.0)	11 (91.7)	15 (100.0)	
Unlikely/extremely unlikely	3 (8.1)	2 (20.0)	1 (8.3)	0 (0.0)	
Body mass index < 18, N (%)					0.91
Extremely likely/likely/neutral	18 (48.6)	5 (50.0)	5 (41.7)	8 (53.3)	
Unlikely/extremely unlikely	19 (51.4)	5 (50.0)	7 (58.3)	7 (46.7)	
Digital ulcers or vascular disease, N (%)					1.00
Extremely likely/likely/neutral	17 (45.9)	5 (50.0)	5 (41.7)	7 (46.7)	
Unlikely/extremely unlikely	20 (54.1)	5 (50.0)	7 (58.3)	8 (53.3)	
Severe pulmonary hypertension, N (%)					0.66
Extremely likely/likely/neutral	33 (89.2)	8 (80.0)	11 (91.7)	14 (93.3)	
Unlikely/extremely unlikely	4 (10.8)	2 (20.0)	1 (8.3)	1 (6.7)	

Outcomes in SSc transplant recipients are reported to be similar to other indications, which could be because of conservative candidate selection in this cohort.[11-14] We conducted this national survey to determine practices of lung transplant centers regarding patients with SSc focusing on consideration for candidacy, pretransplant GI workup, posttransplant management of GI disease, and perceptions regarding posttransplant outcomes. To our knowledge, this is the first survey focusing on aspects of lung transplantation in patients with SSc.

There was a near consensus that SSc is not an absolute contraindication to lung transplant (>90%); despite this, however, only 60% of centers reported transplanting any patients with SSc in the last year. The reason for this discrepancy is unclear, we hypothesize that in addition to rarity

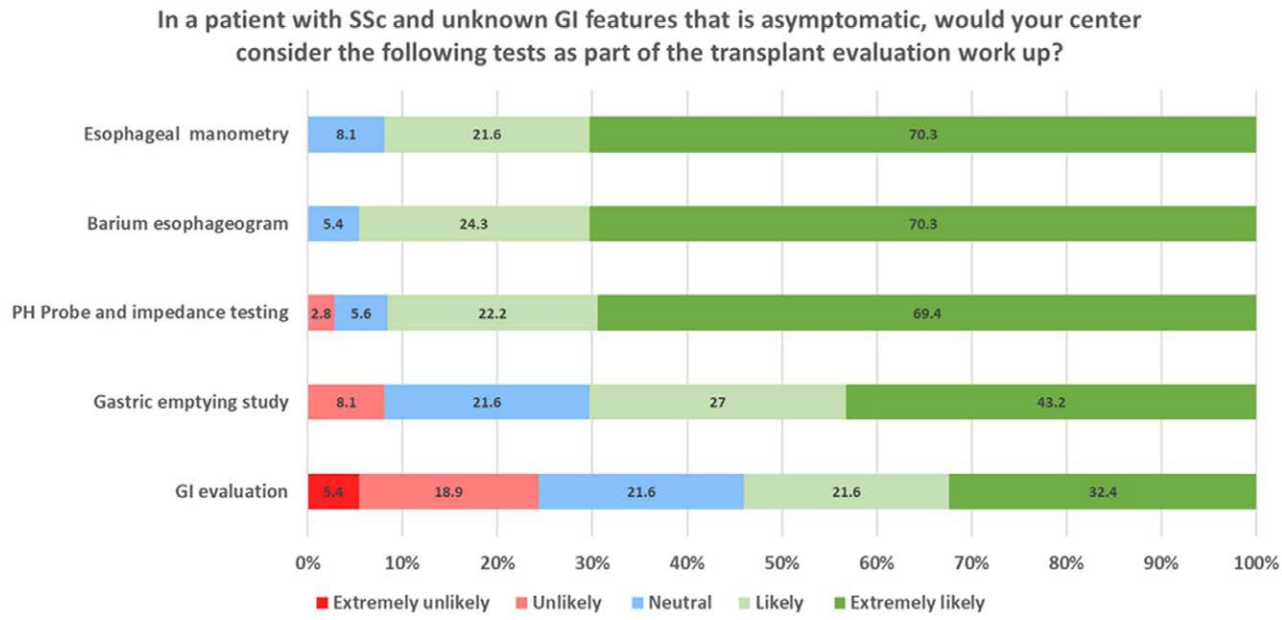

FIGURE 2. Pretransplant workup. GI, gastrointestinal; PH, potential of hydrogen; SSc, systemic sclerosis.

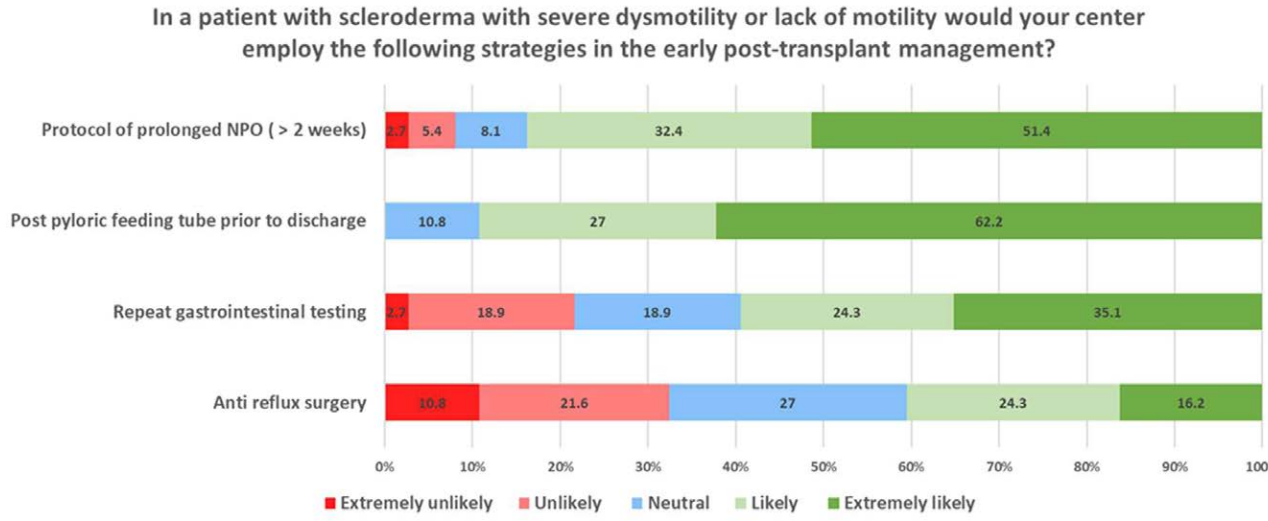

FIGURE 3. Early posttransplant management. NPO, nothing by mouth.

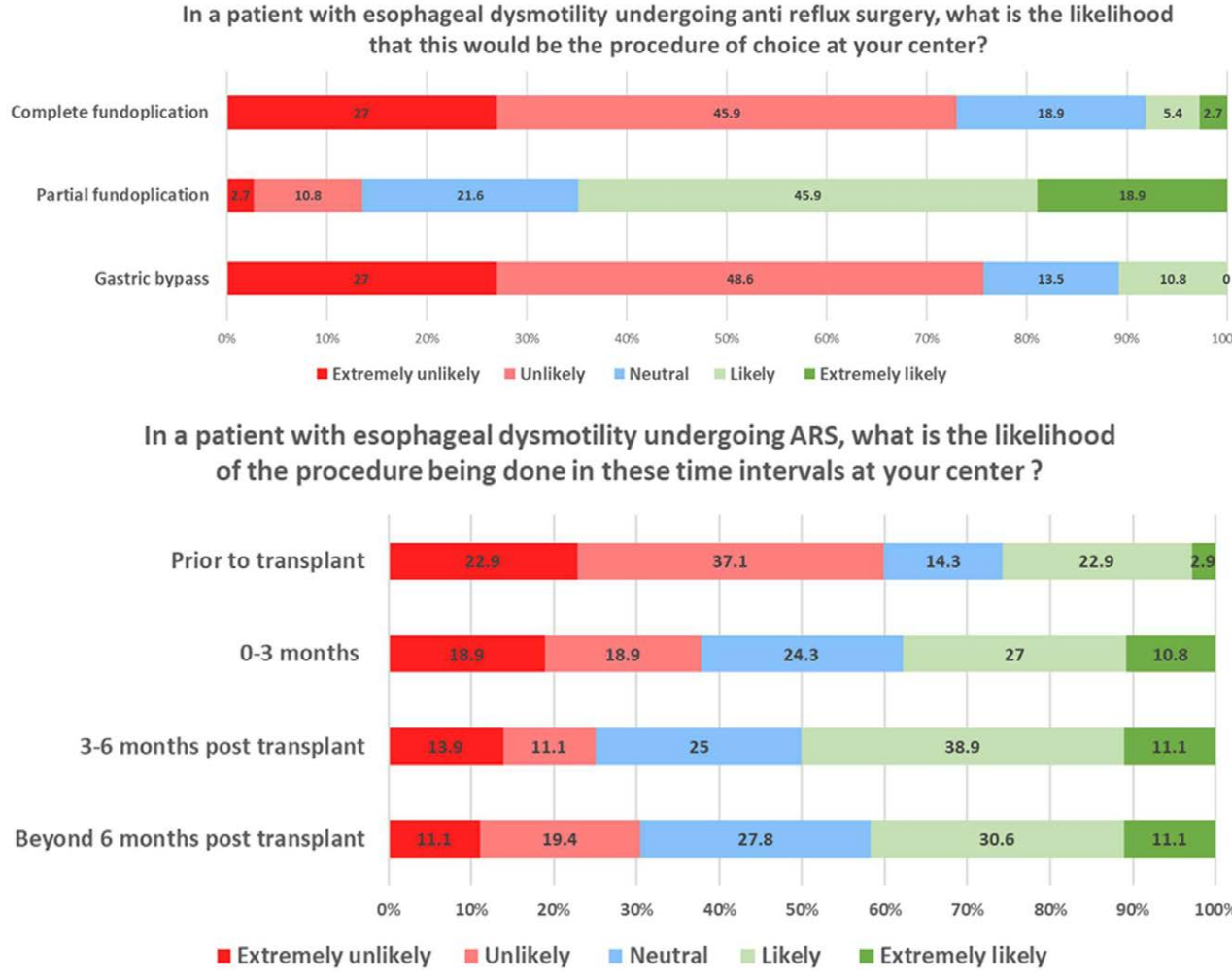

FIGURE 4. Choice and timing of anti reflux surgery. ARS, acute radiation syndrome.

of the disease, it could be because of excessively restrictive criteria used by centers to list patients with SSc. These stringent criteria could also be unnecessarily excluding patients who may have been candidates if not for the diagnosis of SSc. The 3 features most consistently viewed as unfavorable were severe esophageal dysmotility, vascular disease or digital ulcers, and low BMI. Interestingly, most transplant centers did not consider mild to moderate esophageal disease, gastroparesis, or high allosensitization as contraindications to transplant.

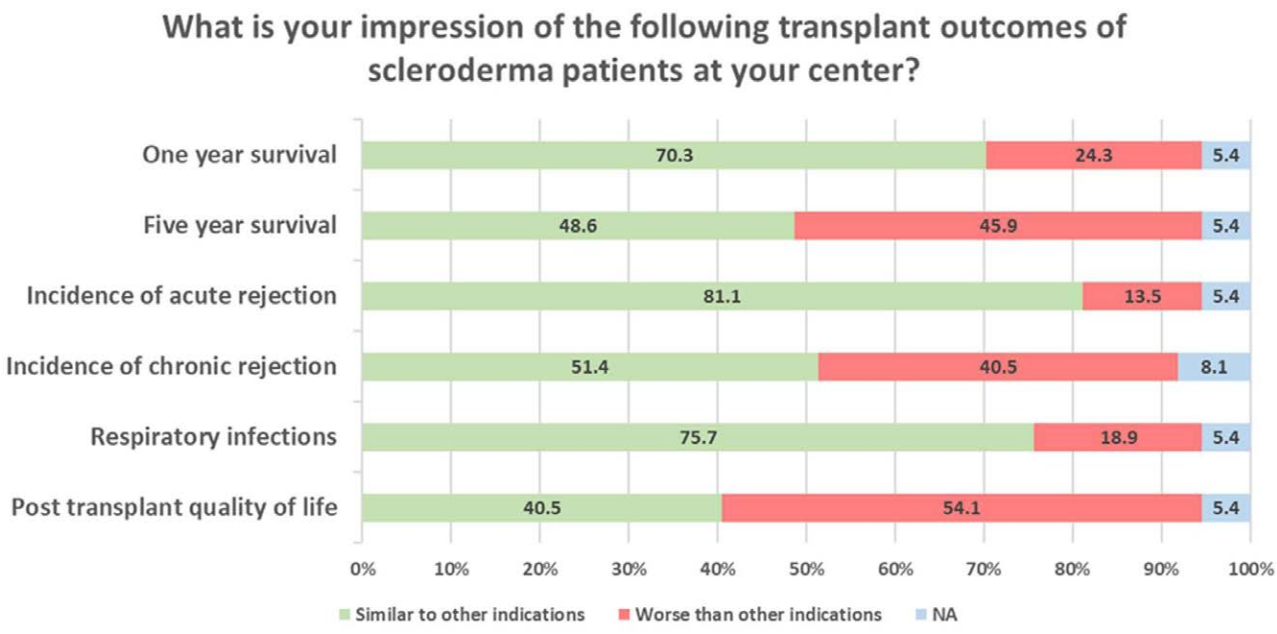

FIGURE 5. Outcomes.

Our survey further focused on workup and management of GI disease since its one of the most common features of SSc and can have a significant impact on lung function. GERD and esophageal disease can often be silent or with minimal symptoms[7]; thus, prompting some experts to recommend screening in asymptomatic individuals.[8,18] The practices based on single-center studies vary widely, with some centers reporting extensive testing for esophageal disease and GERD in all patients[11,12] while others have reported testing only symptomatic patients or those with an abnormal cine esophagogram.[13,14] In this survey, >90% of centers reported doing an extensive evaluation for GERD and esophageal dysmotility using esophageal manometery, potential of hydrogen probe, barium esophagogram in asymptomatic patients with SSc.

GERD and severe esophageal dysmotility in the early posttransplant period can increase the risk of acute rejection, chronic rejection, and impact posttransplant survival.[4,19] There is no consensus in management, with centers reporting a wide variation in practices. Crespo et al[13] and Chan et al[14] reported a conservative approach with prolonged NPO and feeding tube placement, whereas Miele et al[12] and Sottile et al[11] have reported a more liberal approach without a standard NPO or feeding tube protocol. All these studies reported similar outcomes when comparing patients with SSc with other indications at their centers; however, the various approaches have not been compared in any studies. In this survey, a significant majority of centers (>80%) were likely to follow a conservative feeding approach of prolonged NPO and feeding tube before discharge. A smaller percentage were likely to repeat GI testing (59%) or proceed with antireflux surgery (40%). Data supporting a particular approach is lacking; however, practice patterns nationally lean significantly towards the conservative feeding approach.

Antireflux surgery has shown to increase freedom from chronic lung allograft dysfunction and increased survival in patients with GERD.[19] In the presence of esophageal dysmotility, surgery can take additional complexity because of risk of dysphagia and esophageal outlet obstruction. Several reports of fundoplication being safe and effective in the early posttransplant period have been published.[20] Prospective trials of efficacy of reflux surgery in lung transplant patients are lacking. In our survey, the centers were divided regarding the use of antireflux surgery in SSc patients (40%) were likely, and 32% of centers were unlikely to perform. A partial fundoplication was clearly the procedure of choice, and most centers were likely to do the procedure 3 mo or beyond in the posttransplant period. SSc can also lead to oropharyngeal dysphagia, leading to chronic aspiration. This aspect was not addressed in the survey; however, it merits consideration in evaluation of aspiration risk in SSc patients.

The majority of respondents reported that the incidence of acute rejection, infections, chronic rejection, 1-y survival, and 5-y survival to be similar to other indications. These impressions about complications and outcomes are consistent with contemporary evidence wherein most studies report similar outcomes in SSc patients compared with other indications.[1,11] When asked about quality of life, most respondents felt SSc patients had a worse quality of life than other indications. We did not explore the reasons for this impression in the survey. Quality of life posttransplant is impacted by several aspects: chronic rejection, type of transplant, infections, medication adverse effects, and psychosocial factors.[21] Whether additional disease features or management strategies specific to SSc patients impact quality of life posttransplant is not known and should be explored in future studies.

There are several limitations to this study. While the response rate was 60%, we had a broad representation of transplant centers. An inherent limitation of a practice survey is that it is based on perceptions of the respondents and not review of data. However, since most of the respondents were medical or surgical directors, we anticipate that responses were based on substantial knowledge of the program practices and outcomes. The survey assessed the number of SSc patients transplanted at each center but did not address how many patients were evaluated and turned down, thus unable to provide an assessment of restrictiveness of each center. This survey was intentionally

brief to only focus on clinically significant topics, which limited our ability to explore the reasons behind responses. The severity of esophageal disease was not defined in the survey and was a subjective assessment by the respondent. This may impact the generalizability of these responses. Another limitation of the study design is the nonanonymous nature of the survey, wherein responses could be tracked to the respondent. The responses needed to be linked to the center to only use one response from a center, at the same time maximize the number of centers surveyed. The responses were deidentified for analysis, and no survey responses were excluded from the analysis. We recognize that practice of transplant medicine is nuanced, and a brief survey may not do justice to it. However, we believe it provides important insight into the current thought process of transplant programs.

Although published studies have reported varied approaches to SSc patients, we found practice patterns across the United States to be quite consistent. Most programs had similar opinions about favorable and unfavorable characteristics for candidate selection. Extensive pretransplant GI testing and a conservative feeding plan in the early posttransplant period are also widely adapted. However, there was variability in the adoption of antireflux surgery in patients with severe esophageal disease, as well as timing of surgery relative to transplant. In the absence of guidelines for patient selection, information about common practices can be particularly helpful to transplant providers and can start to shape consensus practice allowing for meaningful clinical research. In addition, this knowledge can lead to early intervention for certain extrapulmonary features potentially improving chances for lung transplant candidacy. For GERD and esophageal disease, antireflux surgery can decrease microaspiration and progression of lung disease.[19,20] Digital ulcers may be prevented with lifestyle measures and treated with vasodilators.[22] Malnutrition can be targeted by a multidisciplinary approach to help prevent or reverse weight loss.[9] Future randomized controlled trials should focus on determining best practices for these complex patients; compare management techniques for esophageal disease, timing, and type of antireflux surgery; and measure physiological changes with different treatment protocols on aerodigestive system.

Conclusions

Lung transplantation in patients with SSc has evolved from being considered a contraindication in the early days of lung transplantation to being widely accepted through the United States. Severe esophageal dysmotility, vascular disease, or digital ulcers and low BMI are features that continue to significantly affect lung transplant candidacy of SSc patients. Extensive pretransplant testing and a conservative feeding approach in the early transplant period are widely adapted by programs across the United States. With this study, we were able to shed light on the current practices of transplant centers, although the best practices remain unknown. Future prospective studies comparing different approaches of management may help define the best way to manage patients with SSc who require a lung transplant.

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