December 21, 2021

CTS INSPIRATIONS

CTS NEWS

President's Message

I was reminded by Angela last week that this would be my "last CTS President's message"…. as I prepare to transition to CTS immediate past president, it is truly hard to believe how quickly the year 2021 has passed. It was certainly a busy year, which is probably why it passed in the blink of an eye.

Despite the environment that surrounded us, CTS has made many significant steps made *as we have continued to work to meet the educational needs of our members and ensure the financial health of our organization*. The pandemic has provided unique opportunities to innovate and improve. Most importantly, as we continued to ramp up the momentum of our online webinars with a revised pricing structure more favorable to CTS and free to members, we were able to consolidate to a single in-person annual conference, launch a new CTS **Job Board**, outreach in revised membership tiers, and focus on website redesign.

It was a smooth *take off* as we began the year with Phil Porte, our Executive Director who served CTS successfully for years, who announced his retirement in early spring. Thank you very much Phil for being there for CTS and for ensuring a smooth transition, the entire CTS board appreciates all that you have done over the years. Jason Seidler, our current executive director along with Jade Orchard brings a multidisciplinary skillset to CTS. We are lucky to have Jason take over the reins flawlessly while maintaining the vigor that CTS workflow needs.

It takes a collective effort to make an association like ours successful, and I must give special thanks for all the volunteer members on our Executive Committee, including Michelle Cao (President Elect), George Su (Secretary), Tisha Wang (Treasurer), Laren Tan (Immediate Past President), Nicholas Kolaitas and Ni-Cheng Liang (ATS CCR), as well as the entire Board of Directors. This list is not complete without thanking our conference chairs Gaurav Singh and Kristina Kudelko, and most importantly, I must thank our Education Chair, Shazia Jamil, who has contributed to the success of the CTS conferences for almost a decade. I do know that our entire board is invigorated, motivated and eager to work with the ongoing leadership of the members who remain on the board.

I also want to thank our CTS office staff for making this a very successful year. Vickie Parshall and Dave Eubanks are outstanding representatives for our organization, and with the continued support of Jason's talented team, we only get better. I am certainly not done ☺ and will continue to serve in my role as the Past-President next year, especially to support Michelle Cao, CTS' incoming President. Michelle has been an active board and executive member and will provide excellent leadership as CTS continues to expand its activities and influence. Please join me in welcoming Michelle in her new role.

New officers start their term January 2022. CTS members who have *not yet voted*, kindly cast your valuable vote using the link below. If you know of colleagues that may be interested in joining or participating in CTS, please share our newsletter, website and/or our contacts.

Finally, I thank you all – the members of CTS – for allowing me to serve you as president over the last year. It has been an honor, a joy, and an experience that I will always cherish. I have met many new, interesting colleagues, made many new friends, and enjoyed it to the fullest. I look forward to seeing you at upcoming CTS Monterey meeting this spring. Details of our upcoming conference, scheduled for March 11-13, 2022, will be released shortly.

Sincerely,

Vipul V. Jain, MD, MS
UCSF Fresno

EDITOR'S NOTE

It is one thing to get it, another to keep it, and a third to deserve it.

Addressing Burnout: Moving from Treatment to Prevention

We are so busy trying to "succeed", that we have no time to live.

Working in healthcare has always been fraught. Our compassion and empathy, our caring for the sick and dying, render us vulnerable to emotional distress and fatigue. Even prior to the pandemic, the shift to hospital based care, the creation of the modern ICU, and rise of the EMR further exacerbated stresses by increasing workloads and creating the expectation of 24/7 care. The result is a system driven by productivity metrics where patient care no longer comes first, but competes against the demands of hospitals and insurers leaving us little time for our families and friends, let alone ourselves.

Superimpose the stigmatization, short-staffing, and inadequate personal protective equipment during the initial phases of the COVID outbreak followed by the rancor caused by fake news and misinformation, it's no wonder that clinician burnout has become a *crisis*[1] affecting all members of healthcare teams.

Most readers will be familiar with some if not all of the emotions described in a recent JAMA article including fear, fatigue, isolation and alienation; and betrayal. [2] As Krystal Craddock, CTS' CSRC liaison noted in our OCT 2021 issue, burnout affects not just physicians and nurses but respiratory care therapists (RT) as well.

> "*Of the RT respondents reporting burnout, these individuals worked more hours per week, worked more hours in intensive care unit, reported more exposure to COVID-19, and were more likely to work in community hospitals.*[1] *There were no demonstrated differences in burnout for highest degree earned, role within the department, years as an RT, commute time, shift worked, gender, or race, basically pointing to the fact that, we're all burned out."*[2]

So what can be done and more importantly, what is *being* done? Studies indicate that both individually focused and organizational strategies are needed to prevent and reduce in burnout among health care professionals.[4] As Ms. Craddock further describes in this months's followup article, interventions to address burnout have mostly dealt with individual approaches such as taking time off, exercising, practicing mindfulness, and counseling.[5,6]

While we welcome the increased availability of these resources, it begs the question of institutional accountability in preventing moral injury and mitigating burnout. The CEO of a major academic healthcare system recently wrote that working in healthcare requires passion and sacrifice—but surely not at the *unnecessary* expense of our physical and mental health. As we anticipate yet another variant and another surge[7] this winter, system-level solutions that incorporate the intelligence and knowledge gained from our experience over the past 18 months are urgently needed that not only address the ongoing shortages in staffing, but also re-define FTEs to account for the currently unrecompensed non face to face time spent caring for patients and time needed for recuperation and rejuvenation.

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Connective Tissue Disease-Related Interstitial Lung Disease

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Take-Home Points:

- Interstitial lung disease is a common manifestation of connective tissue diseases and can lead to increased morbidity and mortality
- A timely and comprehensive assessment is important in addition to ruling out drug-related pneumonitis and infection
- A multidisciplinary discussion is recommended in the decision-making for those with ILD.[4] Inclusion of a rheumatologist is crucial in those with suspected CTD-ILD
- Immunosuppressants are the cornerstone of therapy, and antifibrotics could be beneficial in those with progressive fibrosis
- Managing other comorbidities, obtaining appropriate vaccinations, oxygen therapy, pulmonary rehabilitation, and referral to lung transplantation or palliative care (when appropriate) are other important considerations in the care of these patients

Introduction

Connective tissue diseases (CTDs) are a group of systemic autoimmune disorders characterized by immune-mediated injury to many organs and commonly manifests as interstitial lung disease (ILD). In the presence of ILD, patients with CTDs have increased morbidity and mortality. ILD is most often identified in those with an already established CTD. However, in some patients, it could be the initial manifestation of the CTD and therefore patients may present for initial evaluation by a pulmonologist.[1] The prevalence of ILD varies significantly among the CTDs: systemic sclerosis (SSc, 40-80%), rheumatoid arthritis (RA, 10-30%), polymyositis (PM)/dermatomyositis (DM) (40%), Sjögren's syndrome (SS, 40%), and systemic lupus erythematosus (SLE, 8-12%).[1] Patients can also present with ILD with subtle clinical features of autoimmunity without meeting criteria for a definitive CTD. A recent research classification "interstitial pneumonia with autoimmune features (IPAF)[2]" was established to encompass this group. CTD-related ILD is associated with a more favorable prognosis than idiopathic ILDs, therefore, it is important to differentiate the two diseases.[3] A multidisciplinary discussion is recommended in the decision-making for those with ILD.[4] Inclusion of a rheumatologist is crucial in those with suspected CTD-ILD.[5]

Clinical findings

In those without a prior diagnosis of CTD, certain clinical symptoms and signs could suggest an underlying CTD: joint pains/swelling and morning stiffness with RA; sclerodactyly, digital ulcers, Raynaud's phenomenon, telangiectasias, abnormal nailfold capillaries, gastroesophageal reflux disease and dysmotility with SSc; and muscle weakness, Gottron papules, heliotrope rash and mechanic's hands with DM.

Guidelines recommend serological testing to evaluate for CTDs in those with newly identified ILD, however, there is no consensus on the extent of testing that should be performed: RA (RF, anti-CCP), SSc (anti-SCL-70, RNA polymerase-III), SS (anti-Ro/SSA, anti-La/SSB), PM/DM/ Antisynthetase syndrome (anti-Jo1, PL 7, PL 12, EJ, OJ, KS, MDA5, Mi-2), overlap syndrome (anti-PM/SCL), mixed connective tissue disease (anti-RNP), and SLE (dsDNA, Sm).[4]

CT/Histological Patterns

The prevalence of the specific ILD histopathological pattern varies by the underlying CTD. The joint pains/swelling and morning stiffness with RA; sclerodactyly, digital ulcers, Raynaud's phenomenon, telangiectasias, abnormal nailfold capillaries, gastroesophageal reflux disease and dysmotility with SSc; and muscle weakness, Gottron papules, heliotrope rash and mechanic's hands with DM.

NSIP is the most prevalent pattern in all CTD-ILDs except RA, in which UIP is more prevalent. Combined NSIP-OP is seen commonly with inflammatory myositis. LIP pattern is common with SS.[7]

A B C D

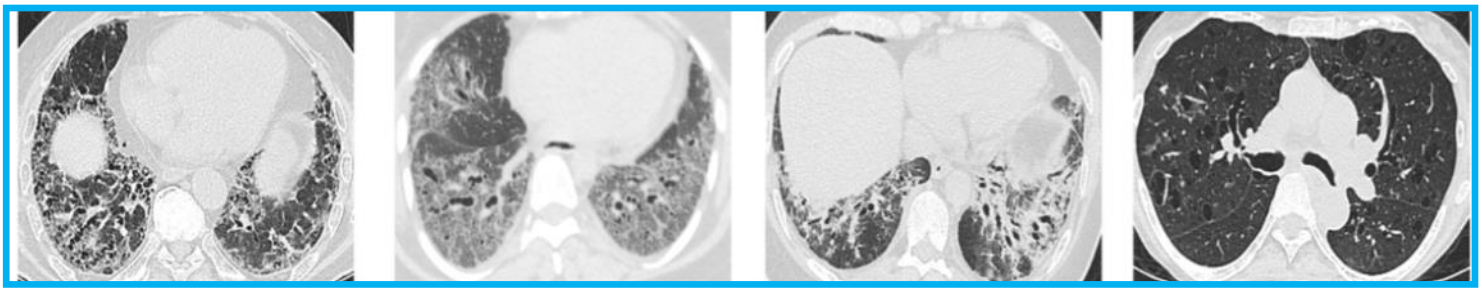

Fig. 1. Patterns of Interstitial Lung Disease on high-resolution computerized tomography. A. Peripheral and basal predominant traction bronchiectasis and honeycombing; usual interstitial pneumonia (UIP). B. Bibasilar ground-glass opacity and traction bronchiectasis; non-specific interstitial pneumonia (NSIP). C. Patchy areas of consolidation in a peribronchiolar distribution, organizing pneumonia (OP). D. Extensive peri-bronchovascular cysts, lymphocytic interstitial pneumonia (LIP).

Lung biopsy is not routinely performed in those with established CTD. However, features on biopsy would include lymphocytic inflammation, prominent lymphoid aggregates with a germinal center, and involvement of multiple compartments.[1]

Treatment

In managing CTD-ILD, it is important to decide when to treat and when to monitor. The evidence to guide clinicians in this decision-making is limited. In one study, a staging system categorizing SSc-ILD as a limited disease (HRCT extent ≤ 20%; or in indeterminate cases, FVC ≥ 70% predicted) or extensive disease (HRCT extent > 20%; or in indeterminate cases, FVC < 70% predicted) at the time of diagnosis was strongly predictive of increased mortality.[8] A significant decline in PFTs (FVC change consisting of either a ≥ 10% decline in FVC or a marginal decline in the FVC with an

associated decline in DLCO of ≥ 15%) during the first year of diagnosis is also highly predictive of increased risk for long-term mortality.[9] The presence of UIP pattern is also likely associated with an increased risk for progression. Treatment should be considered in patients with severe, active, or progressive disease. Other comorbidities and the patient's ability to tolerate the medication should also be taken into consideration.

The current approach to the treatment of CTD-ILD is to select therapies based on the underlying CTD.[7] Immunosuppressive agents are the main first-line treatment and the addition of antifibrotics are considered in those with a progressive fibrotic phenotype. Most of the evidence on the use of immunosuppressive therapy is focused on patients with SSc-ILD. Corticosteroids are commonly used but doses over 15 mg of prednisone equivalent have been associated with renal crisis in those with SSc. In the Scleroderma Lung Study (SLS) I, patients treated with cyclophosphamide (CYC) had a smaller decline in FVC % predicted at 12 months compared to placebo, and the effect was more pronounced in patients with severe fibrosis.[10] In SLS II, mycophenolate mofetil (MMF) was equally effective in improving lung function and was better tolerated when compared to CYC.[11] In a phase 3 trial of patients with diffuse cutaneous SSc, tocilizumab, an interleukin-6 receptor antibody, showed a reduction in FVC decline among patients with evidence of ILD on baseline HRCT (mean change in FVC from baseline at 48 weeks was -14 ml with tocilizumab and -255 ml with placebo).[12] Rituximab, an antibody that targets CD-20-positive B lymphocytes, is another biologic agent that has shown effectiveness in treating SSc-ILD in small randomized studies and observational studies.[7] Autologous hematopoietic stem cell transplantation (SCT) is another treatment option that has shown promise in patients with severe SSc. Compared to CYC, those receiving SCT had improved event-free survival (survival without respiratory, renal, or cardiac failure) and overall survival.[13] Given the increased early treatment-related morbidity and mortality, especially in those with advanced cardiopulmonary and renal disease, these treatments should be limited to specialized centers with an expertise in this procedure.

Antifibrotics, nintedanib and pirfenidone, have also been studied in CTD-ILD. In the SENSCIS trial, in patients with ILD associated with SSc and fibrosis (fibrosis affecting at least 10% of the lungs on HRCT), the annual rate of change in FVC was -52.4 ml per year in the nintedanib group and 93.3 ml per year in the placebo group (p=0.04).[14] In another trial evaluating patients with progressive non-IPF pulmonary fibrosis (26% of patients had CTD-ILD), the annual rate of decline in the FVC was significantly lower in patients receiving nintedanib compared to placebo (-80.8 ml vs. -187.8 ml).[15] In the subgroup analysis, a similar benefit was noted in the subgroup of patients with CTD-ILD treated with nintedanib.[16] In a study of patients with progressive fibrosing unclassifiable ILD (approximately 13% of patients had IPAF), pirfenidone slowed the rate of decline in FVC at 24 weeks compared to placebo measured by on-site spirometry (-17.8 ml vs. -113.0 ml, p=0.002).[17] In another study of patients with progressive fibrotic ILD which included 29% of patients with CTD-ILD, there was a significantly lower decline in FVC % predicted at 48 weeks in the pirfenidone group compared to placebo (although the trial was underpowered).[18]

There have been no large randomized trials published on the treatment of ILDs for other CTDs. For RA-ILD the treatment options are MMF, and in those with significant articular disease, rituximab and abatacept are options. Methotrexate-associated pneumonitis is rare and its influence on progression of ILD was a concern in the past, but accumulating evidence suggests that methotrexate could be protective from developing ILD.[19] The evidence for management of PM/DM-associated ILD is limited. Mild to moderate ILD is treated with corticosteroids and MMF or azathioprine. Patients with severe ILD or refractory to these therapies are treated with cyclophosphamide, rituximab, a calcineurin inhibitor, or tofacitinib (Janus kinase inhibitor) in those with MDA5 positive disease. The evidence for treating ILD associated with SS and SLE is even more limited. Treatment options include corticosteroids, MMF, azathioprine, rituximab and CYC. In mixed connective tissue disease, the treatment choice is based on the overlapping diseases.[7]

Non-pharmacologic Therapies/Comorbidities

Patients with CTD-ILD should also receive other supportive therapies including smoking cessation guidance and appropriate vaccinations. Those with low oxygen saturation levels at rest or with exercise should also be prescribed oxygen. Pulmonary rehabilitation is an important adjunctive therapy to reduce dyspnea and improve both exercise tolerance and quality of life. [20]

Esophageal dysfunction and gastroesophageal reflux are common in those with CTD and could contribute to the progression of ILD. These should be treated with lifestyle modifications and acid-reducing or promotility agents.

Those with CTD-ILD are also at increased risk for pulmonary hypertension (PH) from primary pulmonary vasculopathy and chronic hypoxia. Screening with ECHO should be considered as it may be difficult to differentiate if the respiratory symptoms are due to progression of the ILD, respiratory muscle involvement, or PH. A reduction in DLCO out of proportion to FVC and elevated serum BNP levels would also suggest the presence of PH. Those with elevated pulmonary pressures on ECHO would require a right heart catheterization for confirmation. Those with PH should be evaluated and treated by clinicians with an expertise in this area.

Patients with ILD experience cough, dyspnea, fatigue, anxiety, and depression. Early involvement of palliative care can help with the management of symptom burden and end-of-life care.

Those with progressive disease despite treatment should be referred early for lung transplant evaluation. Lung transplant outcomes for these patients are comparable to those with non-CTD-ILD.[21,22]

Given the complexity of CTD-ILD and the need for a multidisciplinary approach in the diagnosis and management of these patients, referral to an institution with ILD expertise may be beneficial.

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MEET GEORGE SU, MD
CTS SECRETARY

George Su, MD
CTS Secretary
Professor of Clinical Medicine
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What is your story?

My father was the first of seven sons of a part-time military policeman and farmer. My mother was the youngest of seven siblings born to lychee farmers. Both were from Taichung, Taiwan. They immigrated to the U.S. and settled in Stillwater, Oklahoma where my father received his PhD in civil and environmental engineering and my mother received her master's in sociology. I was born in Stillwater as an honorary Oklahoma State University Cowboy birth alumnus. Soon thereafter our family moved to Spring Valley, New York for my father's first post-graduate job at General Foods and then the birth of my sister. Two years later, we moved to Sunnyvale, California for my father's job at Lockheed Missiles and Space Company. Fast forward through piano lessons; perfunctory

Chinese school; acting out Hanuman and Star Trek and spy agent shenanigans with my childhood best friend; camping in Yosemite; soccer, baseball, and water polo; amateur electrical, plumbing, painting, and tiling; Great America games hosting; philosophizing over ping pong with another best friend; losing my mother early to liver cancer; gaining a supportive step mother; dabbling in environmental design and architecture and engineering and then, great fortune had me at the UC Berkeley Department of Biophysics playing with bubble surfactometers and measuring liquid and solid contact angles of lung surfactant components. Foreshadowing indeed. I now serve in a job I love as faculty in the Division of Pulmonary, Critical Care, and Sleep Medicine, UCSF School of Medicine at San Francisco General Hospital. My partner Lisa, a design strategist at ERA-co, and I live in the San Francisco Richmond district with our three children Ethan, Garrett, and Tahlia.

Which is more important: what you say or how you say it?

I think we're all at least loosely familiar with Mehrabian concepts of tone of voice being more effective than word semantics in communicating positive and negative emotions. And now, we're all VERY familiar with the (inappropriately) generalized lore of dichotomizing what and how for all kinds of communication. Nevertheless, since I work in and identify with a profession that values facts, evidence, and truth, I will say that what you say is foundational. Then, if what also includes phrasing, positioning, and framing, I submit that what is more important than how. My truth, however, is a hedge. Impactful communication requires optimization of both what and how. Then, to deconstruct this rhetorical question further, I wonder, as we plummet deeper into communications dominated by voiceless interactions, if what and how will completely cede their importance to who, when, where, and why?

What is a story of yours that you don't get to tell often enough?

I was seven years old and sat by myself on a firm green chair in the corner of a large living room. Silver tinsel chased stuttering lights across the ceiling and up, over, and around a frosted Christmas tree. My father and mother were standing in the center of the room talking to my father's boss and his family. We were at his house. And we were attending his annual Lockheed environmental engineering holiday party. I momentarily lost sight of them between bodies and limbs of the socializing droves. But I could hear them. My father had always spoken loudly and my father's boss' voice boomed beyond. They were talking about a recent project that had gone well. My father's boss announced to the room: "Merry Christmas, everyone! And congratulations, we did it!". He looked over to my father and thumped a heavy hand on his back. "Well done, Su!" My father smiled over horn-rimmed glasses that had slipped to the tip of his nose. His face was still flushed from egg nog sipped nearly an hour before. Then my father's boss leaned towards my father and guffawed: "Su, you did great! Clearly you're smarter than you look!" My father's co-workers laughed in line. My younger sister was startled by the extra noise and buried her face into my mother's dress. Since everyone was laughing, it was all in fun. Right? My father raised his glass of egg nog, without a blink or flinch, straightened his back and bellowed: "Thank you very much for the compliment!" He laughed. But he laughed knowingly.

My father passed away during my pulmonary fellowship. I never did share with him my seven-year-old trope experience. And I never had a chance to thank him.

Addressing Burnout: What Tools and Resources Are There?

By Krystal Craddock MSRC, RRT, RRT-NPS, AE-C, CCM

It has been well established that physician and nursing burnout has been present for several years. The healthcare field is a high stress career and commitment, where all healthcare providers are at risk of burnout. Burnout is characterized by emotional fatigue, depersonalization, and poor efficacy, and it can lead to poor patient outcomes, lapses in professionalism, and a negative effect on healthcare operations.[1,2] This was made further apparent during the COVID pandemic. Studies indicate that both individually focused and organizational strategies can result in reductions in burnout among healthcare providers.[2]

Interventions to address burnout have been identified such as taking time off, exercising, practicing mindfulness, and counseling.[3] One systemic review identified five programs with significant effectiveness on burnout that include Cognitive Behavioral Therapy, meditation, mindfulness, breathing, and relaxation techniques.[4] At the time of this review, they found that seven of these programs were virtually accessible for healthcare workers to easily utilize and access.[4] With the COVID pandemic promoting society to embrace technology for many services, there continues to be growth in access to these programs. Additionally, many of these techniques and tools have become more and more supported and even offered by institutions; however, not all bedside staff may be aware of all the resources.

It is important for institutions and leadership to advertise such resources to alert their teams, to help team members, and to hopefully reduce burnout and turnover. Medical societies can also promote these tools to help members be abreast of the new findings and resources available. There is still much to learn about burnout and resources to help us avoid and/or treat it, but these techniques are a good starting point for all of us. Everyone should look into their employer-offered resources and share them amongst colleagues as well.

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Southwest Journal of Pulmonary and Critical Care, Volume 23, Issue 5 (November 2021)

The November 2021 Case of the Month is a patient with a solitary adenocarcinoma metastasis to the lung who developed lung torsion after resection.
The Medical Image of the Month describes a patient with Pseudomonas pneumonia who developed a cavitating pneumonia. The topic is briefly reviewed.
"Impact of Recent Job Loss on Sleep, Energy Consumption and Diet" discusses job loss effects on sleep, activity and diet.
Lastly, "Rapidly Fatal COVID-19-associated Acute Necrotizing Encephalopathy in a Previously Healthy 26-year-old Man" describes a patient who developed this rare complication of COVID-19.

Volume 23, Issue 5 (November 2021)							
Title (Click on title to open the manuscript)	Journal Section	First Author	Year	Vol	Issue	Pages	Date Posted
Rapidly Fatal COVID-19-associated Acute Necrotizing Encephalopathy in a Previously Healthy 26-year-old Man	Critical Care	Raschke RA	2021	23	5	138-43	11/17/21
Impact of Recent Job Loss on Sleep, Energy Consumption and Diet	Sleep	Batool-Anwar S	2021	23	5	129-37	11/16/21
Medical Image of the Month: Cavitating Pseudomonas aeruginosa Pneumonia	Imaging	Kiladze G	2021	23	5	126-8	11/2/21
November 2021 Imaging Case of the Month: Let's Not Dance the Twist	Imaging	Panse PM	2021	23	5	115-25	11/1/21

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