


Management of Systemic Sclerosis Patients in the COVID-19 Era: The Experience of an Expert Specialist Reference Center

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ABSTRACT

OBJECTIVE: COVID-19 pandemic represents a serious health emergency that severely compromised our Public Health system, resulting in a rapid and forced reorganization and involved the management of chronic diseases too. The Scleroderma Unit of Modena and Reggio Emilia follows more than 600 patients suffering from systemic sclerosis (SSc) and recently became the referral center (HUB) in Emilia-Romagna for this rare connective tissue disease. The aim of the present study was to evaluate the extent by which the lockdown and the pandemic has impacted the activity of admissions to Scleroderma Unit of Modena and Reggio Emilia.

METHODS: Our daily clinical activity is characterized by outpatient visits, videocapillaroscopy exam, ulcers treatment, therapeutic infusions in day hospital regimen, multidisciplinary visits following our dedicated SSc care pathway, and clinical trials. Our activity has been quickly rescheduled to ensure the proper assistance to our SSc patients during the COVID-19 pressure.

RESULTS: The use of telemedicine has certainly assured a robust continuity of health care. Furthermore, telephone pre-triage, nurse/medical triage, proper physical distancing and use of PPE/DPI allowed us to re-organize and continue SSc daily activity. Specifically, therapeutic infusions in day hospital regimen and outpatient visits, including ulcers treatment, was guaranteed and maximized.

CONCLUSION: The management of scleroderma patients by an expert specialist reference center is crucial in order to ensure continuity of care and pursue the best SSc practice.

KEYWORDS: Scleroderma, COVID-19 pandemic, chronic diseases management, remote follow-up

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Introduction

The 2019 coronavirus virus (COVID-19) rapidly raised world concern because of its high contagiousness, morbidity, and mortality.¹

On 11 March 2020, after assessing the levels of spread and severity of the SARS-COV-2 infection, the World Health Organization (WHO) declared that the COVID-19 outbreak recorded over the past months could be characterized as a pandemic.²

Updated data on the epidemiological situation in October 2020 count more than 440 353 023 confirmed cases of COVID-19, including 1 173 777 deaths worldwide.³

Today, COVID-19 represents a serious health emergency that may inevitably involve the management of chronic diseases too.⁴

Systemic sclerosis (SSc) is a chronic systemic autoimmune disease characterized by vascular damage, autoimmunity, and fibrosis.⁵ Currently, we do not know yet the SSc patients susceptibility to SARS-COV-2 and which may be the disease course in those SARS-COV-2 infected.⁶

SSc patients could have a higher infectious risk because of their autoimmune dysregulation and chronic immunosuppressant treatments if compared with the general population. The use of disease-modifying drugs (DMARDs) is associated with a potential increased incidence of infections but an active not controlled disease may be a significant factor favoring infections too.⁷ However, data suggesting that a drug-induced immunosuppressed state might predispose SSc patients to SARS-COV-2 infections are poor.^{6,8} No specific data are available about the risk of a SARS-COV-2 infection in patients previously diagnosed with Interstitial Lung Disease (ILD).

The International Societies of Rheumatology have proposed a set of recommendations to optimize the management of COVID-19 patients and to decrease the risk of infections in rheumatic patients undergoing immunosuppressive therapies.^{9,10} In these patients, a tight follow-up is crucial to verify the specific cases and clinical needs. In particular, the infection risk in SSc might also be related not only to disease activity but also to possible flares due to therapy discontinuation.⁶



While the practitioner should always encourage patients to practice a social and/or physical distancing and appropriate hygiene measures, the reference centers should guarantee a close assistance to SSc patients during COVID-19 outbreak when the routine care activities are upset.¹¹

Scleroderma Unit of Modena and Reggio Emilia

Our Scleroderma Unit follows more than 600 patients suffering from SSc and recently became the referral center (HUB) in Emilia-Romagna for this rare connective tissue disease.¹² Vascular and fibrotic processes resulting in skin fibrosis and multiple organ manifestations, such as interstitial lung disease, pulmonary arterial hypertension, and digital ulcers are frequent manifestations of the disease. SSc treatment requires both immune-modulating therapies and vascular therapies for microangiopathy.¹³⁻¹⁶

Our daily clinical activity is characterized by outpatient visits, videocapillaroscopy exam, ulcers treatment, therapeutic infusions in day hospital regimen (prostanoids, intravenous biologic drugs, and immunoglobulin), multidisciplinary visits following our dedicated SSc care pathway, and clinical trials. Physicians, nurses, specialized in ulcers advanced medications too, and data manager for clinical trials are involved. Autologous stem cell transplantation and challenging studies about stem cells are also performed in our patients with SSc,¹⁷ becoming a therapeutic model with potential socio-economic impact in the national and European network for rare diseases.¹⁸

The aim of the present study was to evaluate the extent by which the lockdown and the pandemic has impacted the activity of admissions to Scleroderma (SSc) Unit of Modena and Reggio Emilia. We considered SSc medical out-patient clinic visits, including digital ulcers care and management, and day hospital admissions for various tasks (intravenous therapies, follow-up visits, and multidisciplinary visit).

SSc Management in the COVID-19 Era

Patient self-care education

A proper self-care education of SSc patients during the SARS-COV-2 outbreak was required. Patients were invited to follow a social and/or physical distancing and appropriate hygiene measures according to WHO and Italian Ministry of Health recommendations.¹⁹ A consistent support regarding working topics was also given especially for SSc patients at moderate and high risk of getting infections in working settings. While over the 80% of economic activities were closed or limited, some of our SSc patients were anyway still employed (medical personnel, ground transportation, etc.) and needed some advices on their risk about infection (based on the available data) or different kind of medical certification (ability/inability, illness, etc.).

According to International Societies of Rheumatology recommendations and the Italian Society of Rheumatology (SIR) about COVID-19 pandemic,^{6,9,20} we explained how to maintain and/or modulate home therapies, even the

immunosuppressive treatments. We highlighted that in SSc patients the infection risk might also be related not only to disease activity but also to possible flares due to therapy discontinuation that should be avoided. Our Hospital Pharmacy and territorial Pharmacies, in accordance with territorial sanitary agencies and/or pharmaceuticals companies, has guaranteed home supplies of drugs.

Scleroderma Unit activity during the lockdown period

A retrospective analysis was conducted in the SSc Unit of Modena and Reggio Emilia

(Emilia-Romagna), regarding all patients admissions during March to May 2020, with special attention to March 9 to May 18, 2020 (lockdown period). Modena and Reggio Emilia provinces showed similar rates of incidence.

Our results were compared with the same period of 2019, without the perception of the existence of COVID-19.

No ethical approval was requested from the different Institutional Review Boards for a simple review of the medical records, since the collection of these data was performed retrospectively but based on data routinely collected during daily clinical practice.

Using diverse ways of communication (video call, skype, mobile phone, email), we guaranteed a solid and daily contact with our SSc patients unable to reach the hospital, guiding them in the management of their disease, with particular attention to treatment issues.

A dedicated SSc nurse, supported by medical opinion where necessary, performed a telephone pre-triage 3 days and 1 day before patients' scheduled visit or therapy/medication. All patients were contacted. During these communications, we assessed possible contacts with COVID-19 patients and/or potential risk of COVID-19, investigating about the occurrence of typical symptoms in the last 15 days. Four patients reported to be infected (one hospitalization without complications), whilst 6 patients reported contacts to SARS-COV-2 infected persons without developing the infection.

A nursing triage was certainly assured at hospitals and department entrances, including Scleroderma Unit and day hospital, asking for symptoms suggestive of COVID-19 and guaranteeing measurement of body temperature, hand disinfection, surgical masks to the patients. The access was forbidden or limited for caregivers.

Doctors, nurses, and all the sanitary equipe wore (and currently wear) the proper personal protective equipment/individual protection devices (PPE/DPI) and applied recommendations to prevent contaminations. Our SSc staff was used to wearing proper individual protections and followed infections surveillance even pre-COVID, in particular during medication procedures.

The outpatient visits and ulcers medications were organized in order to avoid overcrowding as well as day hospital infusions.

Day hospital infusions were re-scheduled respecting the required physical distancing between subjects (at least 2 m).

During the COVID-19 lockdown period (March-May 2020) in Modena and Reggio Emilia we counted 320 total visits of 219 patients at day hospital infusions compared to 335 accesses of 220 patients in 2019 (about 5% less visit than 2019). Mainly prostanoids but also intravenous biologic drugs, and intravenous immunoglobulin were performed according to patients' clinical picture.

Moreover, the service of skin ulcers and wound medications has been maintained in 45 severe patients with 113 total visits in 2020, with respect to 50 subjects treated with medications in the same period of 2019. Therefore, regarding ulcer visits, 90% of the patients who attended ulcer clinic in 2019 came to ulcer treatment in 2020. SSc patients with ulcers who cannot be present in hospital were guided in the correct wound care along with their caregivers or nurse, with the support of calls and photos.

The activity of the SSc medical out-patient of Modena and Reggio Emilia was maintained for severe cases, with the aid of telemedicine for the remaining controls.

Conclusions

The COVID-19 outbreak has severely impacted our Public Health system, resulting in a rapid and forced reorganization.

The organization of the Scleroderma Unit of Modena and Reggio Emilia has been quickly rescheduled to ensure the proper assistance to our SSc patients during the COVID-19 pressure.

The use of telemedicine has certainly assured a robust continuity of health care. Furthermore, telephone pre-triage, nurse/medical triage, proper physical distancing and use of PPE/DPI allowed us to re-organize and continue SSc daily activity.

Huge effort were made to re-organize our scleroderma unit during the pandemic and warrant an almost-normal patient care preserving all our activities (therapeutic infusions in a day hospital regimen, outpatient visits, ulcers treatment/medication), which accounts for about 90% to 95% SSc patients attending to our unit during the lockdown, compared with the same period of previous years.

The management of SSc patients by an expert specialist reference center is crucial in order to ensure continuity of care,

reduce possible complications and pursue the best practice of this rare and harmful disease.

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