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Medical Humanities

# Physical Limitations of Scleroderma- A Personal Perspective

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Scleroderma, like many misunderstood chronic diseases, put a tremendous burden on both the patient and their family. In this deeply personal narrative, the first author describes the impact of scleroderma on her family.

Traveling has always been an integral part of my life. There is just something about experiencing the world with your parents. Exploring Italy and all the rich history it has to offer was a breathtaking perspective; however, it was also a challenge due to the extreme amount of physical energy required. For a normal family, this experience could be easy and filled with much excitement. For mine on the other hand, not so much. The tour to see the Colosseum started near the ruins which meant we had to walk through many obstacles to get to the main attraction. It was a humid day with the temperature being around 90 degrees, making it a difficult walk for anybody. As we walked through, I could see my mother getting tired–her pace slowing rapidly. With every opportunity, she made sure to find a seat before starting her walk again. Our family started to fall behind on the tour and we began to miss key details our tour guide would tell us. The other group members also noticed this and started to stare at our sluggish pace. Part of me felt embarrassed that we were slowing down the group, but the other part felt sorry for my mother who I knew was giving her best effort. I've never felt bad about being an only child, but that day, it felt like the entire world was upon me. It was at that specific moment that I realized exactly how different my family had become over the last three years. This is because of the autoimmune disorder that my mother has.

Scleroderma—something that will always separate us from the normal family.

Scleroderma brings about fibrosis, the thickening and tightening of tissue, and inflammation throughout the body. It can also be divided into two subgroups known as localized and systemic scleroderma. Localized scleroderma involves the skin and subcutaneous tissues, while systemic scleroderma also involves internal organs.[1] This autoimmune disorder is known to be three times more common in females than males and tends to overlap with other similar disorders such as rheumatoid arthritis...which my mother also has.[2] As a person ages the incidence of this disease also increases with most symptoms developing between ages 40 and 50 and a few years after the diagnosis of another autoimmune disorder. It is an extremely rare disease as only 100,000-300,000 people in the US are diagnosed with it.[1] One of the main indicators of being diagnosed with scleroderma is the thickening of the skin which can be extremely painful. This is also accompanied by the Raynaud phenomenon in which the tips of a patient's fingers lose color and turn blue.[2] This phenomenon usually occurs when a patient comes in contact with a cold surface leading to a low oxygen supply from their fingers. Joint pain is also common if the tendons are involved.

Wha is most often noticed with scleroderma is how much a person's physical appearance changes. What many people often fail to understand is how much a person's physical activity changes. Due to the tightening of the skin and inflammation of the tendons, patients tend to have lower grip strength.[3] This also leads to fingers bending forward to the point where it becomes extremely difficult to even make a fist. Because of this, even daily habits and chores become a challenge. Not only do tasks that involve the hands become strenuous but also tasks that involve the legs. For example, a person diagnosed with scleroderma will start to see their pace slowing down.[4] Additionally, knee flexion is reduced, often creating friction and a grating sensation between the tendons.[5] Therefore, when a person with scleroderma chooses to sit down, it takes a while to slowly get back up, especially from seats that are closer to the ground. One of the most life-changing effects of scleroderma; however, is the physical and emotional fatigue it evokes. Fatigue could be a significant source of disability in scleroderma patients as it correlates with pain, longer disease duration, and greater functional limitations.[6] Patients also tend to deal with microstomia, a reduction of the oral opening, which can lead to the rigidness of the tongue and lower oral hygiene in general.[7]

A majority of the treatments used to help accommodate these physical limitations involve motion exercises.[8] Range of motion exercises help to prevent the retraction of the skin and elongate the muscles and tendons.[8] For example, making each finger touch the thumbs helps with movement. To facilitate the flexion of the arms, the hands can be rolled on top of a medium-sized ball. These are two techniques my mother began earlier in her diagnosis and have proven to be extremely beneficial as they allowed her fingers to gain mobility. Another viable treatment option involves the use of heating pads at a temperature of 40 to 45 degrees Celsius.[9] This method can improve the stiffness present in the joints, reducing the difficulty in completing daily tasks. Taking a hot shower and massages can also help to increase the skin temperature and mobilize skin fluids.[9]

To help with the microstomia, one can perform mouth-stretching and oral augmentation exercises.[8] One way to do these exercises is by placing two fingers, one from each hand, into the sides of their mouth which can be done twice a day for 15 minutes each.[8] Another way can be to place a soft wood stick into their mouths at an angle near the molars so the mouth can be kept open.[8] These exercises have improved the mouth opening of patients allowing them to eat, speak, and take better oral hygiene measures. All of these treatments play significant roles in inducing mobility back into a scleroderma patient. As long as one continues with these treatments on a daily basis, they are sure to see progress.

All of these limitations are something that my mother faces on a daily basis. It was difficult to see her change from a healthy, active person to someone who struggles to climb the stairs. From a person who was independent, to someone who now depends on others to complete certain tasks like picking up milk bottles and opening Tupperware lids. Even with something as simple as cooking, she needed someone to assist in stirring and cutting the ingredients on a daily basis. This in turn allowed me to become more independent by learning how to assist in cooking her dishes. Although it felt like I had to be present for every remotely difficult task my mother came across, it allowed me to learn and mature as a person.

This early development of maturity helped me to better understand and support my mom when engaging with the medical community. The world of scleroderma is truly based on trial and error. Scleroderma is still quite unknown to the medical field as it is a very rare autoimmune disorder. Therefore, it was important that my father and I gave her the confidence she needed to help face the uncertainties of this disease. This is also why most physicians are mostly learning alongside the patient. A few months back my mother had her appointment with her rheumatologist and explained how her fingers were starting to bend more causing more stiffness and pain. He suggested that the oral medications were no longer effective, and decided to move on to a different type of treatment that could help with this specific problem: Iloprost and immunoglobulin infusions.[10] According to the rheumatologist, these infusions could allow for the medicine to be absorbed at a faster rate which could relieve the pain more efficiently. My mother and her doctor did not necessarily know if the infusions would positively impact her hands, but this was the best and only option they had at the time. Any effort was better than not trying to improve her discomfort at all. After my mother's infusions started, her hands improved in flexibility and her pain slightly subsided.

Scleroderma is a disease that has forever changed my mother's and by extension my family's life. Similar to any autoimmune disorder, scleroderma is known to be a challenging disease. This is why it was important for my father and I to help my mom feel comfortable without making her feel helpless. We had to be in a position to be able to understand that although she was not able to perform her usual duties, she was trying her best. Due to her fatigue, we had to realize that in some moments she may be overwhelmed which could turn into other powerful emotions such as anger and sadness. My father and I made sure to take all of this into consideration while providing her with the safest environment to get better and heal both physically and mentally. It became important for our family to go on vacations, spend quality time with each other, and have proper conversations to express our feelings. Due to the rarity and lack of knowledge on Scleroderma, it is of the utmost importance to spread awareness and continue to properly research the disorder. In the meantime, my father and I continue to advocate, support, and encourage my mom through her journey, because through perseverance, there are no limitations.

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