Fall 2019

The Hapless State of Amyotrophic Lateral Sclerosis in India: A comprehensive look at life and medical services for ALS patients in rural Himachal Pradesh

Carson J. Bergström
SIT Study Abroad

The Hapless State of Amyotrophic Lateral Sclerosis in India: A comprehensive look at life and medical services for ALS patients in rural Himachal Pradesh

Carson J. Bergström

Pacific Lutheran University, Tacoma, WA, United States

Dr. Sudhir Sharma, M.D.

Indira Gandhi Medical College, Shimla, Himachal Pradesh, India

Dr. Azim Khan, Ph.D.

School for International Training, Delhi, India

Author Note

This study could be conducted thanks to funding and support from the School for International Training.

Table of Contents

Acknowledgments

I would first like to thank the patients, caretakers and healthcare workers that so openly shared their perspectives on their conditions with a stranger. This study would not have been possible without their willingness to talk about their challenges for the betterment of the future. I would also like to acknowledge the help that has been provided by the Indira Gandhi Medical Center in Shimla, Himachal Pradesh. I would like to specifically Dr. Sudhir Sharma, M.D. and his staff for introducing me to and coordinating with these patients and caretakers who have been affected by ALS. I would also like to thank Dr. Azim Khan, Ph.D. for his assistance in crafting this fieldwork study. Abid Siraj also had an instrumental role in coordinating this project. An immense thank you to Goutam Mehr for Indian cultural and Hindi language knowledge that was built in preparation for this study. I would also like to thank Bhavna Singh and Archana Mehr also for their accommodation and Indian cultural teachings to help me with preparation for this study. An enormous thank you also to the Singh family for hosting and helping me in Delhi. This study would not have been possible without the social expertise and care that came from the SIT Public Health program including our dear friends Babu and Champa. This study is dedicated to my grandfather who died of ALS in 2016, my family of dedicated caretakers and those with the disease around the world. May his legacy live on through inspiring further change for those living with ALS to live better lives around the world.

Abstract

Amyotrophic lateral sclerosis (ALS) or Lou Gehrig's disease is a neurodegenerative disorder that leads to paralysis. The disease requires a high amount of medical intervention and interdisciplinary focus to achieve quality of life for patients. This study looks at ALS patient's lifestyles, their access to these medical devices, Indian therapeutic approaches and policy that impacts patients in Shimla, Himachal Pradesh. As caretakers have a critical part in the care for this disease, their lives were also considered in the case studies. It was found that ALS patients are not experiencing access to medical devices necessary for life because of physical accessibility barriers, financial barriers and a lack of knowledge about the disease. Knowledge of government healthcare financial policy and awareness of services offered by Asha Ek Hope to improve the access to these devices for ALS patients was not found in the study. Access to one of two pharmaceuticals was found to be achieved across the study group, however, the treatment methods for edaravone were inaccessible because of physical barriers and finances. With no known cure for the disease, the focus switches towards the Indian perspective on life and offering a death with dignity and comfort. It was found that ALS patients and their caregroups do not support passive euthanasia as determined by a 2018 Supreme Court decision while palliative care was found to be inexistent despite a dire need for this supportive care for families and patients living with ALS. The findings of this study shed light on the dire need for support of these patients in accessing devices and political advocacy. This study also brings public health awareness to the rare disease community in India—breaking down the public health and rare disease oxymoron.

Keywords: ALS, Asha Ek Hope, caregroup

The Hapless State of Amyotrophic Lateral Sclerosis in India: A comprehensive look at life and medical services for ALS patients in rural India

Amyotrophic lateral sclerosis, better known as ALS or Lou Gehrig's disease, is a neurodegenerative disorder causing rapid paralysis of voluntary and involuntary muscles. The disease manifests itself in debilitating symptoms such as inability to move, talk, swallow and eventually breathe. ALS ends up affecting five in every lakh of Indians (Sondhi 2018). Despite a low prevalence, with its poor prognosis, it has been estimated that approximately 1.5 lakh Indians die each year from complications associated with ALS (Sinha 2010). Information about the pathophysiology and pathogenesis of the disease are both unknown and there are currently only two approved etiological pharmaceutical therapies for ALS patients; riluzole and edaravone. These medications have been shown to increase life by only five months. With ALS being a physically debilitating disease with no known cure, the conversation in medicine is more towards supportive care mechanisms. Several different medical devices and procedures are needed to be able to live with the disease. The list depends on the wishes of the caregroup and patients for length and quality of life, however, it can consist of: mobility aids, breathing aids, safety, and cleanliness aids, eating aids and communication aids. Mobility aids include devices such as canes, manual wheelchairs and motorized wheelchairs to aid in mobility for a patient. Breathing aids with ventilators are rarely non-invasive as muscles for breathing are typically affected by the disease. This forces many patients to undergo a tracheostomy to have a mechanical ventilator inserted into the windpipe. Safety and cleanliness aids include bed guards and rail grips to prevent injuries, shower chairs, and catheter insertions are also often needed to prevent discomfort and aid in the disposal of waste. It is also common for ALS patients to undergo a percutaneous endoscopic gastrostomy to insert a feeding tube as eating becomes impossible.

Communication aids are diverse with patients opting to use simple verbal or non-verbal communication, picture cards, or eye-tracking computers for communication. Because of this extensive list of medical devices for people with ALS, there are a list of government programs and non-governmental organizations (NGOs) in developed countries that offer aid to ease the exorbitant financial burden of these devices to families. However, in developing countries, ALS is not taken on as a public health concern because the nature of the disease's treatment presents a challenge to a developing healthcare system. Curing the disease is impossible, the treatment methods are non-traditional and its prevalence designates it as a rare disease. This, coupled with no cure for the disease, leads to a lack of political incentive to help these patients. In the Indian context, ALS devices for patients are unknown because government healthcare policy surrounding rare disease to support costs of medical device procurement has been placed in abeyance and moved into a national healthcare scheme logistically incapable of covering the disease. To make life more complicated for ALS patients in India, there is a lack of neurologists in rural India. Most Indian neurologists reside in larger cities and India's rural, underdeveloped and economically challenged population is suffering greatly from this misdistribution of specialists (Gourie-Devi 2008). Quality of life is important to consider in a well-rounded discussion of ALS in any country and choosing to live may not always be an option for patients. With recent developments in political policy surrounding death with dignity practices such as passive euthanasia and palliative care, ALS patients were included in policy as groups who have the right to undergo these options for death with dignity. The status and opinion of this practice's use by physicians and patients with ALS are currently unknown in the country. This paper will investigate first-hand accounts from patients, caregroups and healthcare professionals in rural parts of India related to healthcare surrounding ALS. It mainly aims to communicate the

caregroup's accessibility to medicine, the need for comprehensive care and health policy for ALS patients in India and other challenges the community and other rare disease patients in India are experiencing. Solutions and political advocacy efforts to the problems being experienced will also be proposed following interview data analysis.

Literature Review

In 2018, a study with thirty-two people living with ALS at the Indira Gandhi Medical College in Shimla, Himachal Pradesh was published that surveyed demographic information about the population (Sondhi 2018). This study is a rare publication, showing for the first time in the 21st century, clinical information on patients who have successfully navigated a referral process in the Indian healthcare system to bring their condition to the tertiary level of a neurologist. Important information on the course of the disease in India was discovered as the disease was found to take longer to progress with a mean time for limb and bulbar onset of 19.06 months (Sondhi 2018). This study showed male preponderance was not a factor in Shimla as has been seen in other places (Sondhi 2018). It also confirmed similar study findings when compared to Western countries there is a younger age of onset and longer prognosis of life for Indian ALS patients (Sondhi 2018). This finding that the disease is occurring at a younger age and with people living longer, created an urgent concern over quality of life and access to medicine and proper care for these patients which was the primary focus of this paper.

ALS is a disease in one of its most vicious incarnations. Many who are diagnosed with the disease experience an incredible sense of urgency to scramble and cling to the rails of anything resembling hope. Many find this hope in the ways of traditional and more alternative forms of medicine. In a recent study published by Srivastava from Deharadun, Uttrakhand, hope is illuminated for patients through the practice of Ayurveda. Srivastava reported a case of ALS

that was treated with panchkarma that took an ALS patient's ALSFRS-R, or rating of ability from the disease, and improved it by five points (Gusain 2018). This is a feat unheard of in modern medicine and may be inspiring patients to begin to switch to Ayurveda. Connections with Ayurveda extend into offering hope as well for a sudden cure. There is a connection to the town of Shimla and the story of Hanuman's effort to treat Lakshmana which was done with an Ayurveda treatment of a believed neurological disease cure-all herb, *Sanjeevani booti*. With Ayurveda also being central in faith for some patients in India, this study also sought to understand if patients following diagnosis by an allopathic doctor, seek alternative forms of medicine as the options in allopathy are very limited for treatment and offer little hope for a cure.

Another study published by Thomas later in 2018 on the effects ALS has on caretakers, found the financial burden of the disease to be an immense strain on caretakers (Thomas 2018). This study also highlighted the importance of governmental policies, because, in 2017, there was a recent policy meant to curb these financial strains by implementing the central Indian government's National Policy on the Treatment of Rare Disease (NPTRD). This policy would completely cover the cost of medical devices for ALS patients. This policy was not working as of 2017 though, as only 180 applicants applied for support. However, the policy was yet to truly be tested as no one ended up being accepted and six pediatric patients died waiting for funding on the list (NORD Report 2018, Sinha 2010). In 2018, the next year, the policy was placed in abeyance to redefine rare disease outlined in the policy. These patients were put into the Ayushman Bharat scheme, commonly referred to as Modicare, however, there are potential barriers to this scheme. Modicare is not as comprehensive in providing devices and care as the rare disease policy would have been. This study observed what changes have occurred with ALS patient's access to healthcare and medical devices without this policy's implementation. It also

looked at any knowledge that the community has of the ALS NGO, Asha Ek Hope that is operating out of Mumbai. Understanding the interdisciplinary study of politics when considering the health of ALS patients is incredibly crucial as government policy and healthcare implementation of quality of life measures are also an instrumental part of living with ALS.

Currently, with the disease's poor prognosis, quality of life is an important question to ask within ALS caregroups. Quality of life can be defined in many ways, however, in this context, quality of life shall be defined as a set standard of health, comfort, fulfillment, and happiness defined by individuals or the group affected. One way to measure quality of life is through the evaluation of an individual's desire to choose to die. A study done in 2017 examined ALS patient death wishes in India through the course of their diseases through "Wish to Die Questionnaires" (Gourie-Devi 2017). The overall findings of the study found that these patients were in psychological states where they were still positive about life and found life worth living. A limitation of this study was that many patients were earlier on in the disease and still reported independence as only four out of the twenty reported being dependent on others. To offer another conclusion to the paper, it may have been the continuity of autonomy into their cases of ALS that has led to patients seeking to prolong their lives. The loss of autonomy is the most difficult part of the illness for patients and what has driven patients in developed countries to seek out euthanasia (Wang 2016). Later stages of the disease are incredibly debilitating and this leads many patients to choose a death with dignity. Passive euthanasia was legalized through a judgment made by the Indian Supreme Court in 2018 despite the Indian constitution specifically outlawing suicide. The policy set up and implementation are yet to be evaluated and there is yet to be a study done on terminally ill patients and their caregroup's perspectives of this change. This study particularly focused on conversations with patients, physicians, and caretakers who

have both witnessed and experienced the loss of autonomy through ALS and their opinions of the practice. This study did not only look at interviews from ALS caregroups but also a lawyer from the capital of Delhi who is educated on the Supreme Court case that ended up legalizing the practice–giving important information on the origins of this policy for the country.

There is a contrast with the definite stand that the Indian Supreme Court decision on the legalization of passive euthanasia as a form of death with dignity and the Indian government's lack of commitment in supportive death through palliative care. As of the time of this study, the national government has yet to make a policy on palliative care in the country. Health is a state concern in India, however, this has opened an opportunity for inconsistencies across the country in palliative care. There are currently only two states in India that have any policy on palliative care, Kerala and Karnataka (Khosla 2012). Domestically, in India, there is a great model for palliative care in the Indian context. The state of Kerala has become a World Health Organization model for offering patients low cost, high-quality palliative care that has led to patients experiencing higher quality of life (Khosla 2012). The state of Maharashtra has drafted a policy on palliative care, however, it is still yet to be implemented (Khosla 2012). As this policy is developing in Maharashtra, it is unknown what healthcare professionals in the state are feeling with the potential of palliative care as a policy in the state or in a state like Himachal Pradesh that currently does not discuss policy regarding palliative care. Furthermore, there are hurdles in implicit bias patients may be experiencing with beliefs that Indian patients experience less pain than other ethnic backgrounds which may lead to less focus on pain management palliative care practices (Siong 2019). This study will talk to ALS patient's caretakers about their feelings on palliative care and their desires in Himachal Pradesh to see policy implementation. This study also investigated first-hand accounts from healthcare providers from the state of Maharashtra in

India on their perspectives of palliative care and its implementation in a state considering policy on the practice.

Methods of Study

Because of the qualitative nature of this study, to get an in-depth look at healthcare and medical devices for ALS patients in India, this study interviewed a diversity of avenues for ALS care in the country. The case studies were based out of the Indira Gandhi Medical College (IGMC) which is based in Shimla, Himachal Pradesh. Shimla is a rural, Western Himalayan town with a population around 1 lakh. The focus of these interviews was designed to uncover barriers in access to healthcare and medical devices for these patients, challenges that are persisting within this small study group and the journey through the Indian rural healthcare system for a patient living with a rare disease to end up being diagnosed with ALS at a tertiary, medical college hospital in rural India. Also, this study asked questions to shed light on the developing palliative care system in India and death with dignity in the country because of the 2018 Supreme Court decision to permit passive euthanasia. This study also tailored the questions to shed light on accessibility issues not only within caretaking and the home but in general life as these patients experience many different levels of ability throughout the course of ALS. Finally, this observational case studied the awareness of Indian governmental policies that may have assisted in care for ALS. It also asked about any awareness people have of the Indian ALS NGO, Asha Ek Hope, and directly focused on the NGO remotely providing patients medical devices and medication. Verbal consent was obtained for each interviewee of this study. Questions for this study were developed carefully with the collaboration of other experts not to cause emotional distress for the interviewee. This study and the proposal for funding was approved by a Local Review Board organized by the School for International Training based in Delhi, India.

Living with ALS in Shimla, Himachal Pradesh

Stephen Hawking, the famous physicist who lived with ALS for over fifty years, is often looked at by the ALS community as an icon of living with the disease. Hawking studied the cosmos and the origins of life for his years as one of the most influential physicists of our time. Hawking once talked about his disease in an interview by saying: "I have been lucky, that my condition has progressed more slowly than is often the case. But it shows that one need not lose hope." ALS patients often want nothing more than to have Hawking's life with the disease. The unfortunate reality of ALS in India is that the hope of living with the disease is not the case even if it progresses slowly. These patients cannot have an opportunity to impact the world because they do not have access to medical devices that Hawking had to prolong their lives. The conversations with caregroups pointed to one thing being the number one concern of theirs—hope. Many patients found their hope in Hindu spiritual methods but their hope is not parallel in the healthcare system as there is no financial or physical way for these patients to live with the disease in India.

In rural Himachal Pradesh, an interview was conducted with a physician who often works with ALS patients, Physician C. This conversation was mostly directed towards what can be expected from a physician during the disease for people in the area and trends that they see with service requests by patients. Life with ALS starts for patients at a stage way before diagnosis. Physician C commented that the disease in patients, "…usually presents with weakness. Maybe in upper limbs, lower limbs, or difficulty in swallowing or speaking difficulties." These differences in symptoms often lead patients to navigate a myriad of different paths within a complicated referral system to see a neurologist for diagnosis and treatment. Physician C also mentioned the importance that this disease needs to have a caretaker that is knowledgeable about

motor neuron disease so that the patient may be taken to a healthcare center for these issues to be addressed by a physician. Most of the time it is just luck that a caretaker is well educated about the nature of a disease like ALS. Physician C mentioned the reality of this disease and the hopelessness of it will lead many to feel like they have no function in society. Physician C mentioned that most patients in rural India will experience a "life without a purpose." They went on to mention that most of these patients, while they do not experience physical pain, they will have to deal with immense dependability. This leads many of these patients in the area to experience end of life neglect as caretaker education about the disease is weak, especially amongst the elderly and those within low-socioeconomic standing. End of life neglect can also be the result of the patient's inability to communicate needs and wants in life because of a lack of access to a method of conversation with their caretakers (Physician C, personal communication, November 16, 2019). The physician experience and knowledge gave initial information on what life with ALS can be like in the area. Conversations with patients and caretakers offered a more in-depth look at the devastating toll this disease can have on a patient in rural India.

In another interview with a caretaker from rural Himachal Pradesh, Caretaker A, the experience of a patient, Patient A, who had passed away was described and the impact that it had on the caretaker's life. ALS is a disease that requires full dependence on a caretaker, this disease was mentioned as being incredibly taxing on the caretaker regardless of the good socioeconomic standing of the caregroup. This patient was fully covered by their caretaker's government health plan that allows patients to have most costs of ALS be reimbursed by the government when the patient used government facilities. This was mentioned to be incredibly important in not experiencing financial difficulty with the disease Caretaker A explained. Despite having great financial coverage of the disease, life with ALS still moved very fast and the patient only lived

for a little under a year despite some of the best care in India. This patient spent a total of thirty-three days in the government hospital at IGMC on incubation. This was a mentally taxing issue for the rotating group of caretakers, however, it did not lead to any financial issues because it was fully covered by the government. After the tracheostomy and throughout incubation in the hospital, Caretaker A mentioned that life began to become difficult since they thought this visit was just a routine hospital checkup of a symptom and doctors ended up putting her on life support and a ventilator hours into the visit. As they walked into the hospital room, seeing their mother hooked up to different tubes and devices made the whole caregroup uncomfortable and had long term psychological impacts. Following the incubation, the patient received a portable ventilator which was noted as one of the only costs that came out of pocket for Caretaker A. The patient regained sensation and was sent back home. The most difficult part for caretakers of the disease was not the physical responsibilities of caretaking, it was noted to be the psychological aspects. Caretaking became difficult because the most common thing asked of Caretaker A by Patient A was to take them outside and to keep them entertained. Caretaker A was quoted saying "Every time the question was the same; when are you taking me outside? How many times can you say the same thing? Wait for two days, wait for two days? Every day was the same. You have to lie." This part of the interview showed the lesser-acknowledged psychological effects of ALS on the caregroup. When a patient is trapped inside of their body, there is a desire of the caretaker to relieve their pain and help them achieve a quality of life. However, there is no possible way to do this in the rural western Himalaya Indian context. Lying about the future of the condition was noted to stress this group of caretakers and creates stresses and moral dilemmas for the caretakers. Lying to the patient was also a common theme throughout the interviews. Another unique aspect to the rural context of ALS is that many caretakers of the

disease, according to Physician C, because of the complex nature and the horrible reality of the progression of the disease, resort to keeping the patient in the blind. There is a lack of education about basic human anatomy and physiology in the rural Himachal area, which means that education on what ALS is, is complicated and impossible in some cases without this basic background knowledge of nerves and muscles. Being in the blind, while it may have benefits for patients to be unaware of the bleak future they will experience with the disease, this conversation opens the door for a myriad of morality issues to arise about the caretaker and patient interaction in any disease and individual patient's rights to know about their health.

With ventilator machines and wheelchairs and an inaccessible world surrounding rural India, this leads many patients feeling trapped as Physician C also mentioned during our interview. Many of these rural Himachal towns are built on mountains and the vertical nature of a city like Shimla offers incredible difficulty for accessibility issues which was also echoed throughout our conversations. This issue of accessibility has been a great concern for all patients with differing abilities, but especially amongst patients living with ALS. To get up to the main area in the city of Shimla from the bottom of the mountain, you have to take two lifts, where the entrance is guarded by a flight of stairs and no ramp. Then to be able to access the main point of the city from the lifts, there is a hill that has an incline of about 10 degrees at a minimum for a kilometer or two. To say that the city is inaccessible for patients with ALS or other neuromuscular diseases is an understatement. Physicians and caretakers agreed that more needs to be done in cities to improve accessibility for patients including in their homes. In the situation of Patient A, she was forced to walk down to the primary health center to receive edaravone medication for her condition. The hill in which she lived on though, was so steep that it was impossible throughout her disease for her to walk down, receive treatment for hours through an

IV and then walk back up the hill. Responsibility went back on the caretaker who then would go and drive to pick up their mother and take her to her house. It was too much of a hassle for the caregroup so the caregroup opted to stop treatment (Caretaker A personal communication, November 18, 2019). It is uncommon for accessibility to be considered during the construction of PHC's in India. Some believe that it is only the tertiary hospitals that are doing these complicated treatments for these rare diseases, however, as shown in this case study, there are healthcare professionals in primary health centers working with neuromuscular patients who are disabled from their diseases, and accessibility must be considered on all levels of healthcare facility construction and renovations in the future. One thing that was mentioned by Caretaker A as being a solution to life with accessibility issues are temples in rural India. Generally, temples in India are made to be as accessible and flat as possible. Caretaker A mentioned that during the disease, they would take their mother out to the local temples as it offered the ability for hundreds of meters of land unobstructed by hills and stairs and a place of worship and hope for their mother. (Caretaker A personal communication, November 18, 2019).

The focus on temples was not only because of accessibility, but it was also because of the importance of Hanuman and the Hindu stories for ALS patients. It was also commented by Physician C that most ALS patients he sees show devotion to Hanuman (Physician C personal communication, December 6, 2019). Caretaker A specifically mentioned visiting the temple because of the accessibility of the temple and being able to freely roam around and worship. Lord Hanuman has incredible importance to ALS patients as the story of Hanuman and Lakshmana tells of an herb that can cure all neurological illnesses that are certain to cause death, such as ALS. This was noted as a piece of hope for the caregroup as they were caring for Patient A (Caretaker A, personal communication November 18, 2019). This unique aspect of the Indian

experience with ALS shines a light on the more interdisciplinary importance of caring for patients and caretakers of the disease. It also shows that the spirituality that was observed of ALS patients in this study may not only be due to accessibility of temples but also because of the spiritual importance the stories can provide patients as well.

The chances of a Stephen Hawking scenario occurring in India is next to nothing as these patients are being bedridden. The life of an ALS patient in rural Himachal Pradesh is short, reliant on caretakers and locks patients in their homes because of infrastructure issues in India. It was mentioned by Physician C that these people often die waiting for a miracle (Physician C, personal communication December 6, 2019). These patients often do not know what the course of disease will be or that they are hit with the disease in the first place. Their spiritual lives have been noted to be stronger mainly because they have no other places to access in their areas than the temples and devotion to Hanuman specifically may bring hope in a possible cure. There are plenty of societal factors that are leading to the difficult life that these patients are living. However, the lack of access to medical devices to overcome the physical boundaries from society plays a part in not providing these patients the ability to be independent or to even live.

ALS Patient's Access to Medical Devices in Shimla, Himachal Pradesh

Although a wheelchair is a simple device, wheelchairs for ALS patients can produce life-changing effects for patients. Wheelchairs give ALS patients the ability to regain a sense of freedom to move around as they please. Without the ability to walk, many people with ALS rely on these devices for their mobility with the help of a caretaker or, in best-case scenarios, to be independent in their movement through technologically intensive wheelchairs. An issue in India, however, is wheelchair accessibility and efficiency of the device for patients living in rural areas of India. With manual wheelchairs costing around ₹5000 apiece, Physician C explained that there

is an issue for some people affording the service in the first place. Furthermore, complications arise with ALS patients when the disease progresses into the arms rendering most patients to become dependent on a caretaker for mobility. Another option exists in motorized wheelchairs that read patient's eyes to move the chair around, however, that is not reasonable technology for patients who are having a difficult time affording a wheelchair as the price for such a wheelchair in India is around ₹2,00,000. Physician C further explained that within the area of rural Himachal Pradesh, even if people can afford them "most patients do not have wheelchairs. In this area, I have not seen this. Most patients are carried by their caretakers around." (Physician C personal communication, November 16, 2019). This was true when we spoke to caretakers and patients as well, despite the financial or health insurance status of the patient in the case study. When we spoke with Caretaker A from the area, they said that their position at the hospital directly led to their access to a wheelchair for their house. Even with a wheelchair, the patient was still confined to the house after their hospitalization and never left the house up to death (Caretaker A personal communication, November 18, 2019). While this study did show an example of a patient who could gain access to a wheelchair in India through insurance, employment and finances, this example still found that owning a wheelchair was not enough to roam outside because the cities and roads are not accessible for anyone to access as mentioned previously in the *Life with ALS* section. There are not only issues with ALS Indian patients gaining access and being able to afford mobility devices to benefit their quality of life, there are also bigger issues for patients being able to access life-sustaining medical innovations such as ventilators as well.

Patients with ALS, as has been mentioned during the late stages of the disease, require at home, portable ventilation at some point to keep the patient alive. The cost of a portable ventilator was estimated to be ₹2,18,000, or almost 50% of a family's health insurance coverage

from Modicare in India for those not supported by the NGO (Physician C, personal communication, November 16, 2019). It also needs a professional to manage the technology which can vary greatly in the cost between patients. This immense burden and fast progression of the disease then can force those patients out of range, or unaware of the NGO's services and in lower socioeconomic standings to not receive a ventilator, a crucial piece of technology in order to live with the disease. Although Patient A had received a ventilator as told by Caretaker A in the interview, it was noted to be the most financially draining portion of caretaking for the disease and the caretaker had to go to Chandigarh, a town over three hours away from Shimla to procure the device (Caretaker A personal communication, November 18, 2019). Speculation is that patients who are even on Modicare still would struggle to afford the device, service, and upkeep of the device as twenty-four-hour care from a knowledgeable healthcare worker is required to live on a ventilator. This means that ALS patients in India are dying of early-onset, preventable breathing complications unless they have the fortune of good insurance and procurement of a ventilator such as the case of Patient A. These patients are undergoing a death that is completely preventable with the use of modern medical technology. This technology is not being accessed by patients around India because of the high cost of the device, lack of knowledge and the lack of comprehensive medical insurance coverage for medical devices. While it may be hard to attain a ventilator, pharmaceuticals were found to be easier to get, however, treatment for the disease is still expensive despite the inexpensive Indian pharmaceutical market.

As has been mentioned previously, there is no known etiological, allopathic cure for ALS now. However, the two approved therapies that have been shown to slow down the progression of ALS, are; riluzole and edaravone. One promising feature that was mentioned by the caretaker

and the physician is that riluzole is offered by the company Sun Pharma at no cost to patients. This is great news; however, this drug was approved by the FDA in the 1990s for the treatment of ALS and has shown incredibly minimal results in patients.

Most get approximately five more months out of the drug, however, even that is currently being debated and Caretaker A said they did not think riluzole did anything for Patient A (Caretaker A personal communication, November 18, 2019). Another drug, edaravone, was approved in the year 2017 for usage, however, Physician C mentioned that they do not believe that any significant differences have been made with the regimen of edaravone. In a conversation with Physician C, they mentioned, “the patient must come in for 14 days to the hospital, then go home for 14 days and repeat this process for six months.” This treatment regimen is not sustainable or worth it for many patients as the reality of the disease is that many patients will die in the year that Physician C sees them as the referral process takes a long time to reach a diagnosis of ALS (Physician C personal communication, November 16, 2019). Even in the case of Patient A, they would walk down a hill to the local PHC to receive their IV edaravone treatment. However, this was not possible to sustain throughout their life and ultimately, they ended up stopping the treatment due to accessibility concerns despite being able to afford it from insurance. It is important to note that Physician C quoted the treatment at costing a total of ₹51,000 for the complete treatment not including the costs of hospitalization if it is done in a different facility than theirs (Physician C personal communication, November 16, 2019). In a country that champions cheap therapeutics, this therapy is not going to be accessible for most Indians to just be able to afford the medication much less the personnel to carry out the therapy. The issue of affording personnel to help with care for the disease is not limited to edaravone and is also seen in at-home care as well.

The problem of ALS patient access to medicine also becomes a healthcare personnel problem when patients begin to discuss at-home care for their family members. In conversations that we had with Caretaker A and healthcare workers in Himachal Pradesh, there seemed to be a clear distinctive difference between the at-home-care received by those who had comprehensive health insurance because of their job, and those who do not have comprehensive health insurance or health insurance at all. Caretaker A mentioned that to help them with day to day activities around the house and caring for their mother, they hired a full-time nurse to facilitate the care. These duties were mentioned to include tasks such as clearing saliva from Patient A and ensuring that the ventilator was working correctly and in one case, the nurse saved Patient A's life when oxygen levels decreased and the nurse could get access to an additional oxygen source for the patient from the hospital. This would not happen with someone who could not afford the extra care needed to live with the disease. However, Caretaker A explained that this was one of the most expensive parts of the disease and although most other ALS related costs were covered by governmental health insurance for their employees, this one was not and the family was left to foot the bill for in-home care and salary for a full-time nurse from the town of Chandigarh, over three hours away (Caretaker A personal communication, November 18, 2019). This at-home-care is not a reality for many patients in India. Most patients in India will be diagnosed with ALS at IGMC and they will more than likely never return to the center and will die with the disease at their homes within a year from preventable causes of death (Physician C personal communication, November 16, 2019).

Another device that is not being utilized in India very often because of the short lifespans these patients experience is the procedure of feeding tubes. The case study reported showed no issues with swallowing food but a decreased appetite with the disease, which is a sign that the

muscles are deteriorating as eating becomes more difficult, but there was no feeding tube insertion throughout the course of the disease (Caretaker A personal communication, November 18, 2019). Physician C commented on the observed phenomenon as being an issue about lifespan rather than if their muscles are degrading or not. Most patients will not live long enough for swallowing muscles to degrade completely, which leads to patients not ever needing to implant a feeding tube. Although it was mentioned in the interview that percutaneous endoscopic gastrostomies can be offered by IGMC, it is rarely carried out because patients with issues swallowing and bulbar onset of the disease typically live another month as reported by Physician C (Physician C personal communication, November 16, 2019). Overall, as other procedures and devices become more accessible, it will be important that coverage for this disease is proactive in including coverage of feeding tubes as more patients will be able to live longer when devices for living with earlier stages of the disease are covered. When patients can afford to have wheelchairs and ventilators with a world that is accessible to them, then they will be able to advocate for further services to be able to truly live with their different ability level.

This unfortunate reality of living with ALS in rural India is filled with macabre and the reality that a soon approaching death by not being able to breathe is destined for those diagnosed with the disease and many patients do not even know that death is approaching. ALS in rural Northern India has appeared to be defined by patients and caretaker's inability to access medical devices that are necessary to live and live a quality life due to their financial backgrounds and familial vocational pursuits.

The Process of ALS Healthcare Navigation, Diagnosis, and Alternative Therapies

An overarching goal of this study was to use the case study of ALS to begin to start conversations on the experience of a rare disease patients in Indian healthcare. This study

documented the experiences that patients and physicians have had in diagnosing and treating ALS as one example of rare disease in the country. From conversations with Physician C and caregroups who had navigated the system, it appeared that it is not a simple task.

While discussing the process of ALS diagnosis at IGMC, Physician C mentioned that the procedure to get diagnosed with ALS includes a minimum of three visits with various physicians. Physician C commented that as most patients will experience symptoms of ALS that are unusual for regular diseases, most patients will first go to see their local physician in primary health centers for symptomatic reports before getting referred to IGMC for their symptoms. Physician C does not see patients without an in-house referral. Physician C said during the interview about when they come to the IGMC as; "Here they are seen by them presenting symptoms. If they have weakness, they will go to medicine department. If they have difficulty swallowing they will go to ENT or gastroenterologist. And then from there, they are referred to us because we do not take patients directly." (Physician C personal communication, November 16, 2019). This navigation process could end up taking a few different trips to the hospital for a patient to receive a diagnosis of ALS. This time could be crucial time lost as therapeutics are most effective in the early stages of the disease (Brooks 2018).

Once the patient makes it to the neurologist, there is a very limited amount of testing that can conclude that a patient has ALS. Physician C said the diagnosis process takes three to five days while staying overnight in the hospital. During that time, Physician C stated that the process of diagnosis is not able to diagnose ALS, it is mainly able to rule out other diseases. Physician C commented; "The main aim of our workup is to rule out other causes. So that we may not miss something treatable." (Physician C personal communication, November 16, 2019). Once biomarkers show that a patient does not have another disease, then patients will undergo

electromyography (EMG) to test the activity of the motor neurons. EMG is one of the most common ways to diagnose ALS—it is a relatively painful diagnostic test as an electrode must be inserted into the patient's arm for testing. The test lasts an hour and gives results on the movement of muscles following electrical stimulation of motor neurons. This test, along with symptoms of ALS, and lab work showing no signs of other diseases, will lead to a patient's diagnosis of ALS at IGMC. Often, Physician C said that some patients, if they are financially able, will go to another private clinic to confirm or refute the diagnosis, however, they mentioned that the diagnosis is rarely debated because, in India, doctors will determine a patient's fate without question. In the case of Caretaker and Patient A, their coverage allowed them to consult with doctors at the All India Institute of Medical Science (AIIMS) to confirm the diagnosis. This caretaker considered themselves lucky though to have had a physician to diagnose them with their disease as they mentioned inconsistencies across cases to have a physician that will diagnose and then follow up with ALS treatment in India.

Conversations with patients made it appear that these patients were very lucky to have a neurologist in the area and to have one that is committed to caring for them. This was mentioned to not always be the case in ALS for some patients. Some governmental hospitals were told to be refusing to see patients if they have suspected ALS. Caretaker A discussed this in the interview by saying, "Some government hospitals will not advise anything with ALS. If a patient comes to them with MND, they will simply say this is a terminal disease, they will not prescribe the medication, we do not want to waste the money and time." (Caretaker A, personal communication, November 18, 2019). While it is a difficult disease to care for, the healthcare system is required to offer therapeutics and devices that allow patients to live with the disease. Especially patients need to be seen should medical innovation be possible such as the case of

ALS. The lack of doctors willing to help people live productive lives with ALS is hindering ALS patient's quality of life and access to medical devices as well in India.

As some doctors are not being willing to treat ALS, this has forced many patients, as well as the limitations of allopathic and modern medicine's ability to traditionally treat them, to seek out alternative styles of medicine. This is a common trait to see in patients and Physician C mentioned that they recommend alternative treatment in these situations. During our interview, Physician C said "Treatment is limited. It is very common amongst patients to seek out other medicine. I recommend that they seek alternative medicine. Ayurveda has shown some promise with treating patients… It gives them hope if nothing else." This switch of medicine is one of the only alternatives for patients to look to in times of fear. Doctors like Physician C must see the value in giving patients hope if nothing else and Ayurveda helps guide ALS patients through their diseases and their faiths as mentioned through Hindu philosophy and beliefs.

While the treatment of disease in India is typically confined to allopathic and traditional medicine with prescriptions of medications, ALS yet again breaks the mold of common disease and requires rehabilitation for patients to successfully live with their disease. During a conversation with Physician C, they recommended visiting the Indian Association for Muscular Dystrophy (IAMD) center in Solan, Himachal Pradesh, two hours from Shimla to hear about their take on treating and working with patients with muscular dystrophy and motor neuron disease. The progression of muscular dystrophy mirrors some of the progression of ALS as well—which has led to ALS often being connected to muscular dystrophy advocacy organizations and cases in the past. During an interview with the founders and current leaders of the therapy center, they mentioned the importance's physically, mentally and socially of their model of care. The organization is focused on providing patients the ability to live with the

disease with the reality that it is an incurable illness. They have worked with patients with MND and mentioned that they had a patient with ALS come in from Punjab the other month. They can work with patients to help them find medical devices and services through government schemes and NGOs. They mentioned that they also give tips on how to make their living arrangements more accommodating to their conditions. The work IAMD can do should act as a model for future neuromuscular clinic development in rural and urban India. Their work consists of week-long therapy camps that help patients combat the symptoms of muscular dystrophy and teaches people how to live with the disease once they return to their homes in rural and urban settings with great success. The organization also subsidizes the stay for those who cannot afford it and they offer devices to patients who cannot afford it. All the directors being affected by muscular dystrophy themselves, they emphasized the importance that people have access to motorized wheelchairs and them being a way to gain independence. They have advocated strongly for the inclusion of accessibility provisions in political legislation and have had success in their state and the national parliament as well. They mentioned in the conversations that they felt that conducting scientific research in their facility will give patients hope for a cure and hope is a very powerful thing for patients. They also commented that the social aspects of these diseases are often most difficult for these patients. They mentioned that in rural communities it is not common for patients with ALS or muscular dystrophy to leave home and enjoy their lives because the villages will critique their situation telling them that they do not have anything to smile about with this disease. They also mentioned that many pediatric patients that they have will look to end their lives through euthanasia as the disease is socially taxing and places a burden on parents as caretakers. They spoke of their plans for expansion to cover more patients throughout India as they already have two other daycare facilities in Chandigarh and Delhi. Their

approach to treating the disease is social which was noted as one of the most mentioned aspects in our interviews with people affiliated with ALS care (IAMD, personal communication December 4, 2019).

The work that has been done by Physician C, IAMD and caretakers are efforts that deserve incredible praise as their care has not been met by patients being able to financially afford to live with the disease. Their care can only go so far in helping people live productive lives with ALS if a commitment is upheld by organizations to make the process for diagnosis and treatment as efficient as possible as living with the disease needs early education and intervention as the progression of the disease can make learning how to live with the disease near impossible.

Indian Philosophy, Death with Dignity and Palliative Care across India

Atul Gawande, the author of *Being Mortal*, wrote in his book: "Our ultimate goal, after all, is not a good death but a good life to the very end." Gawande's book tells of the reality we all face in life with death as an imminent event. Despite the commonality of death across humanity, we fail to discuss it out of fear which leads to a lot of unnecessary suffering and inconsistent governmental policy. It is ironic though, that in the conversations of quality of life we turn to improve health practices of death. However, to offer a more comprehensive form of patient care in the case of incurable and terminal illness to offer quality of life to the end—this is a necessary conversation to have. This project found, based on first-hand accounts from patients and healthcare providers that the state of palliative care in India needs improvement for patients but death with dignity practice in India lack support within the healthcare community and this needs input from Indian patients to guide further discussions around the practice in the country.

As mentioned previously, Indian policy on death with dignity practice has been highlighted by the Supreme Court's decision in 2018 to legalize passive euthanasia. This practice was passed through the case of Aruna Shunberg when the Supreme Court specified two scenarios in which a patient can willfully undergo passive euthanasia. These two conditions are the brain dead where a ventilator can be switched off and those in a persistent vegetative state where nourishment can be decreased and palliative care based techniques can be applied following international standards. The second part of the ruling directly applies to caregroups and physicians to fulfill the wishes of patients in cases of ALS to undergo passive euthanasia by switching off a ventilator. In an interview with Lawyer A from the nation's capital in Delhi, they mentioned that the main groups advocating to the Supreme Court for this case were human rights advocates that believed that the government could end suffering with terminally ill patients. They also mentioned that they felt that physicians would be happy to hear the results of this case as it now offered doctors ethical guidelines to when to offer these services (Lawyer A, personal communication, October 28, 2019). However, conversations with healthcare providers, caregivers, and patients found that the Supreme Court's decision to allow passive euthanasia has not translated into a patient desire to access or support the service. The decision also has not influenced healthcare providers to facilitate the service. Furthermore, there is also no current policy on palliative care set forth by the national government to uphold the second part of this ruling and palliative care is inconsistent across the states. These factors lead to a decrease in usage of death with dignity and a lack of knowledge and usage of palliative care procedures by patients and healthcare providers as shown in this study.

Consideration of the input of Indian culture and philosophy of life is an important consideration when discussing death and passive euthanasia. In an interview with a physician in

rural Himachal Pradesh, Physician C, they explained the importance of cultural consideration in passive euthanasia. It became clear throughout the study that the Supreme Court's decision has not translated into people moving forward with passive euthanasia even in the late stages of their disease. These findings are coupled with positive outlooks on life, reaffirming Gourie-Devi's previous findings in 2017 (Gourie-Devi 2017). Physician C, as a specialist, often works with incurable diseases where most people are given limited time to live. They commented on a question about passive euthanasia by directly saying, "…euthanasia is an alienated concept [for Indians]. In the sense that our philosophy says that we purify ourselves through suffering." Physician C said that they have never heard of any cases in their career of a patient wanting and carrying out passive euthanasia. As a physician who works with many different rare disease patients, this was a telling sign that there has not been a desire from their patients to undergo passive euthanasia. Specifically talking about ALS, Physician C commented that they do not even have the option of euthanasia as their disease progresses so quickly. Physician C said that patients who come in with bulbar onset of the disease, they typically give the patient a month to live which does not allow ALS patients to even think about passive euthanasia (Physician C, personal communication, November 16, 2019). This hesitation to passive euthanasia is also deeply rooted in the religiosity that ALS patients have as well.

In Shimla, there is an incredibly important monument and temple atop the Jakhu hill to the Hindu god, Hanuman. Caretaker A identified as Hindu and to visit the temple of Hanuman. Hanuman may have importance in philosophy for caregroups and it is believed that belief in him indirectly may lead to the retention of hope in the lives of ALS caregroups in the area. The story that is local to Shimla residents claims that Hanuman rested atop Jakhu hill while on a quest to retrieve a powerful medicinal herb, *Sanjeevani booti.* Lord Hanuman was told to find this herb in

the Himalayas by a priest to cure an injury Lakshmana had sustained while in battle. *Sanjeevani booti* is important to ALS caregroups because it is believed that this herbal remedy is located nearby and can cure any neurological illness that has assured certain death—such is the case of ALS. This retention of a belief that ALS can be cured if this is found is even mentioned by leading physicians practicing Ayurveda in the area (Yadav 2017). Through this belief being central to many caregroup's practice of Hinduism, it makes understanding out of their belief in hope and disagreement with passive euthanasia by holding out for a cure.

Passive euthanasia, while still being a fresh ruling in 2018 has also had a difficult time getting the support of the Indian healthcare culture. Through studies of physician and nurse's opinions of the practice from rural communities within the states of Maharashtra and Himachal Pradesh, there was a consensus amongst those interviewed that the practice will not be conducted under their supervision. Three out of three healthcare professionals in rural Maharashtra and a physician in rural Himachal Pradesh all said that they would not facilitate the practice because they did not feel comfortable with it. During a conversation with Physician A, they mentioned their experience caring for a patient with rabies in another hospital where the other doctor carried out a dose of insulin to end the suffering. However, even with immense suffering being displayed in front of them, they still said that they would not have made that decision to end the patient's life. Physician A went on to explain that there was a need for extreme caution when talking about passive euthanasia. This extreme caution is not only needed to provide the patient with the highest quality of care, physicians need to express caution because it may lead to future legal and potentially physical altercations because nothing is hidden in India. When an unfortunate death happens at the hands of a physician, the blame will always return to the doctor. Fear of backlash may be preventing physicians from being comfortable exercising the practice. In another

interview done with a nurse in Maharashtra, there was found to be continued skepticism throughout the healthcare system on passive euthanasia. From Nurse A's perspective, if a patient is chronically ill and not able to be sustained in India, it was their opinion that a healthcare professional's job is to do everything possible to keep patients alive. They also included that they do not feel it is right to pull the switch on a patient. This nurse also said that no hospitals that they have worked in within Maharashtra will do this practice. The only situation in which Nurse A felt that this would be okay would be if someone was given hours to live, they were in immense pain and the patient was able to give consent to ending their life. These conversations built up the case against the practice of passive euthanasia for any patient in the places that were surveyed during this study. This indicates a need for further conversation in healthcare about the practice and whether to continue the policy if it cannot be supported by physicians. With Indian societal and healthcare cultures seeming to not be agreeing with patients who are in vegetative states being able to undergo euthanasia, there is a need for palliative care to provide quality of life for patients in their death.

As has been mentioned previously, palliative care is a process offered by many healthcare professionals to help make people comfortable and improve their quality of life during the end stages of their lives. There is an ethical obligation to ensure quality of life is carried out for everyone to death. As a part of the journey with ALS, many patients due to its rapid progression and debilitating nature, need to consider supportive, palliative care as a part of their treatment. In conversation with a rural physician in Himachal Pradesh, Physician C felt very strongly that the field of palliative and supportive care needs to be addressed in India, especially for ALS patients. They mentioned during the interview that although they believe patients may not be experiencing excruciating pain during their illness, they talked about life with ALS as, "… life without a

purpose." (Physician C, personal communication November 16, 2019). This study opened the conversation of palliative care to a larger cohort of physicians across specialties, out of ALS and Himachal Pradesh and into Maharashtra.

The people interviewed in healthcare offered a diversity of perspectives on the topic of palliative care—from physicians (Physician A and B) that may suggest palliative care, a nurse that may provide palliative care (Nurse A) and a social worker (Social Worker A) who may work with a family during times of mourning. In an interview with Physician A, palliative care was defined as caring for a condition that is beyond active medicine. This is slightly different from Nurse A's opinion that palliative care's definition should be centered around providing a peaceful death for people. This was also subtly different from Social Worker A's definition of palliative care as a religious and spiritual experience for the patient and family members. Difficulty in understanding the definition and the importance of palliative care may be preventing its implementation in Indian healthcare. In conversation with a social worker from the district, they stressed the importance of palliative care as being a social necessity for closure and important in preventing mental health concerns in the future. The nurse mentioned that they felt that there are more pressing issues in India to take on, such as communicable diseases, than providing palliative care and death with dignity to people. This thinking is part of a more systematic problem with rare diseases and will be addressed later in the paper. This contrast of opinion appears to also mirror the Indian government's commitment to palliative care. The government and healthcare culture in India is still concerned with keeping people alive through any methods possible. This leads to the government not being open to creating a policy to provide comfort to those who are dying. Another problem for patients to receive access to palliative care is implicit bias within Indian healthcare culture with Indian patients. In a

conversation with Physician B, they mentioned that they believe that Indians have a higher tolerance of pain so palliative care practices are not necessary and most people will be okay dying at home without pain mediation procedures such as the administration of morphine. As was shown in another study mentioned previously, this information has been refuted by scientific literature. This belief held may be keeping the people from receiving palliative and supportive care in India. Not only are patients attempting to overcome the government's lack of attention to the need for palliative care, but physicians may have a built-in implicit bias that may be preventing them from achieving a peaceful and painless death.

Death in India is not simple and ALS offers reason for change in the process of death in India. As patients are diagnosed with a terminal disease with no hope for a cure, in Western culture, the patient will have the final opinion in how they will pass away and where their possessions and life's work will go towards. In India, however, that was not found to be the case. During an interview with Physician C, they mentioned that ALS patients, as they have mentioned are in the dark about their condition and typically, they have no idea of the fact that death is soon approaching. It is also uncommon for the caregivers to be consoled during the passing of their family member which leads to emotional complications. Furthermore, the situation becomes even more complicated when the patient has no will to express where possessions or earnings will go to when they pass. The establishment of wills and workers to help families in times of mourning for death in India would greatly benefit life for the patients. Although there is already a positive outlook on life identified by this study and Gourie-Devi, this may enhance that positive outlook for patients with security for their families in the way that they see fit and defining the exact moment in their disease which they do not want to live longer. Currently, these decisions lie in the hands of the caretakers which was noted to cause some strain in

families hit with these types of disease. Death counseling was a suggestion made by Physician C since they said that keeping patients in the dark about their condition may be causing unknown suffering and them not living the rest of the life the way that they would want to (Physician C, personal communication December 6, 2019). This is important to include death counseling and will development in any policy relating to ALS care in the future for India.

Overall, India is running into roadblocks in palliative care and death counseling offerings while passive euthanasia is not being supported amongst ALS patients because of Indian philosophy, Hindu beliefs and their positive outlook on life. This positive outlook on life is optimistic for this study because this still means that without access to devices to help with living life, they are still living a quality life. With accessibility, medical device access, and death practices established as valid concerns for ALS in the country, we turn to changes in policies and organizations that may be able to help the community live better in the future.

Asha Ek Hope NGO Efficiency in Rural India & Shimla, Himachal Pradesh

While there is little to no medical treatment for ALS and access to these medical devices are poor, this has inspired the creation of the non-governmental organization (NGO), Asha Ek Hope. Dr. Hermangi Sane, MD, started and acts as the director of the organization, inspired by her own life living with motor neuron disease. One of the ways that Asha Ek Hope looks to overcome the illness is through a program that gives this medical equipment to patients. This program is a rollover, free-of-cost program for patients to be given these devices to live. The equipment device list is diverse and extensive including ventilators, bi-pap machines, O_2 machines, motorized and manual wheelchairs, as well as braces for limbs. However, as reported, the caretaker, organization and healthcare professionals we had done case studies say that these devices are not present amongst patients nor have they heard of this organization. In every single

interview in Himachal Pradesh, there was no knowledge of the NGO or the services that were offered. This led our group to spread awareness of the organization with the people we spoke with and to reach out to the NGO for an interview on the problem of awareness and the equipment rollover program. However, over the course of repeated communication efforts with the organization, we were not able to reach them, unfortunately. This study hopes that the NGO will gain more awareness and be advertised to more ALS patients through this paper. However, the reason why patients are not using the services could not be identified without speaking with the NGO directly.

Efficiency of Related Governmental Policy in Shimla, Himachal Pradesh

With health being a power relinquished to the states in India, it is rare for intervention to occur on the highest level of the central government. In the case of rare disease, it also means there can be inconsistencies across the country for which patients can be able to afford care. NPTRD looked to resolve these inconsistencies by implementing a program where patients could have associated costs of living with a rare condition completely covered. This policy's implementation in 2017 was abruptly halted in 2018 when the government placed the policy into abeyance and moved all rare disease patients under the coverage of the Ayushman Bharat, or Modicare, scheme regardless of the financial status of the family. Modicare allows for these families to spend ₹5,00,000 on healthcare-associated expenditures at hospitals of their choosing for ₹5 a year. Logistically, this is not enough to care for ALS patients though. This was shown by the study and ALS care for one patient alone easily can reach over the family limit in one year. Issues have seriously developed in the implementation of the policy as no member of the study done in Himachal Pradesh had even heard of the 2017 policy or its abeyance.

During the first seventeen months of NPTRD, only 180 applications were received, none were approved and six children died waiting for treatment regimens to be approved by the government through the policy (NORD Report 2018). The implementation and awareness of government programs to alleviate the financial burden of rare disease was consistent with our fieldwork. During interviews with caretakers and even healthcare professionals, no one had any knowledge of the 2017 policy, the policy's abeyance, or even coverage rare disease patients can get through Modicare or elsewhere. This is incredibly concerning as patients, unless they are employed by the government as Caretaker A was, are covering the entire cost of the disease out of pocket. Even the portable ventilator for Caretaker A was not covered under their insurance, leading to a costly two-lakh price tag that placed a high burden on the caretaker's family. Policy should address the issue of rare disease in the country as so many people are suffering with these conditions in India. These policy changes also need to be inclusive of cases like ALS that will allow for adequate funding for ALS patients to be able to receive medication, medical devices and cover the salary of caretakers that are necessary for providing educated life-saving services for patients. There needs to not only be efforts in political conversations to emphasize rare disease healthcare, but there also needs to be a thoughtful effort on the implementation and advertisement of the services by the government, healthcare workers and other support systems for patients.

There is a reason for optimism though, as a recent coverage for rare disease patients is currently being studied and was advertised through our conversations with IAMD. They mentioned that there is a scheme being rolled out by the Indian government soon, the Rashtriya Arogya Nidhi (RAN) scheme. It identifies certain diseases as life-threatening and offers 2 lakhs to patients to help afford the costs of living with that disease and receiving treatment in

government hospitals. IAMD was confident that this will help patients with muscular dystrophy and even ALS more in the future should it be advertised correctly (IAMD personal communication December 4, 2019). While reading the document released by the government on RAN, while ALS is not specifically mentioned in the study, it does mention that the Technical Committee can consider cases for financial assistance which should grant ALS patients the ability to have medical devices and pharmaceutical therapies such as edaravone covered if the patient desires. It is also unclear if this can be stacked on top of movement into Modicare under the 2018 abeyance of NPTRD. There is further advocacy needed from the rare disease community on how to best treat these patients.

Conclusions & Limitations of the Study

Overall, this study shows what living with ALS may be like in rural Northern India through interviews with a variety of stakeholders in treating ALS patients. The study showed that most caregroups are comprised of close and extended family members and in cases of good insurance standing, nurses who can care twenty-four hours with the disease. It was also able to find that the education of caretakers of those with the disease is incredibly crucial to the outcomes of quality of life with the disease. It is incredibly important because those with the disease are often put in the dark about their condition leaving caretakers to care for the disease without the input of the patients. There is also an increase in spirituality amongst those affected by ALS in the area as temples are common places that people will go with ALS patients, however, this is also connected with the inaccessibility of any other place in the cities of rural India. In the case of Shimla, devotion to Hanuman can also relate to belief and hope in Ayurveda for a cure for the disease. Patients are often confined to lives at home as there is an

interconnection between accessibility in the area not allowing them to be able to go outside and social pressures from communities that shame people living with ALS from enjoying life.

Talking more about the medical aspects of ALS, there is poor access to medical devices and interventions for these patients. This study uncovered that the inaccessibility of healthcare was due to a mélange of different features. Wheelchairs are not being accessed in the area because of the inability to use a wheelchair first and foremost. However, the ability to use a motorized wheelchair is being hindered by not having the funds to pursue it. There is no portable ventilator access for most patients because there is a high cost of the device including someone professionally trained to monitor the functioning of the device. There is little access to people to care for patients twenty-four hours a day, especially in rural areas, and access to it may be divided across who has better insurance for the cost. One positive was that riluzole was found to be free for patients seeking this pharmaceutical treatment, however, edaravone was not found to be accessible for patients because of the high cost and inconvenience of having to come in so often to have the treatment administered by a professional in a clinic setting for weeks on end.

Before receiving a diagnosis, this case study serves as a greater example of what rare disease patients may be experiencing in the healthcare system. It was found that there is an extended and complicated referral process that patients must go through before seeing a neurologist for the condition. The disease may get stuck at treating symptoms of the disease at many different points. Once a patient reaches a neurologist, it was found that modern science can be inconclusive of the disease of the patient, however, they can rule out other treatable cases that present like ALS. There was also mention that there is also a scarcity of neurologists in India that was backed up by data and physicians that will treat ALS is also limited as some see it as a waste of their time. It is common for patients to switch to Ayurveda for their medicine following

diagnosis from an allopathic physician as Ayurveda offers relief in some of the symptoms, but also Hindu beliefs and philosophy may hold powerful hopes for a cure in ALS. It was also found that rehabilitation procedures to help patients live with ALS are being underutilized by patients in the area probably due to inaccessibility concerns.

Death with dignity views and the status of palliative care were also studied as patients are not being offered suitable conditions to be able to choose to live. It was found that ALS patients, caretakers and healthcare workers do not support passive euthanasia and will not conduct the practice out of Indian philosophical values to them. There are also misconceptions and lack of consensus of definition and importance around palliative care and a lack of guiding principles from healthcare in India on what palliative care is and how to provide the service to patients.

Furthermore, this study showed no knowledge of the NGO, Asha Ek Hope and their services that are being offered. The study tried to contact Asha Ek Hope to discuss access to their rural patients in Northern India to give them support through communication languages adapted to ALS, extensive medical devices and offering them community, however, those efforts were not successful and the study is limited to the survey of stakeholders outside of NGOs in ALS care.

In terms of governmental policy, knowledge of any related schemes or insurance developments for patients with ALS was inexistent by those involved in ALS care. Reinstatement of the National Policy of the Treatment of Rare Disease, Rashtriya Arogya Nidhi and Modicare were identified as future policies to advocate for ALS patient inclusion in provisions for funding to receive access to medical devices, death counseling services and at-home care for patients covered by the government to relieve the gargantuan cost of living with ALS in India.

This study did come with limitations—mostly by the case study approach giving such a limited view of what ALS looks like for patients. The limitation of the insurance policy in the case study was noted to not be a common case. This patient was connected to another who had most of the coverage of their costs of ALS under their government employment status. This study was also limited to a public-sector study and the limited sample size may not be representative of the whole community as patients and caregroups may also be seeking a completely private mode of healthcare for ALS. Without talking with these constituencies there is hesitation in applying this experience pan-India with the people living with ALS. We ask for those who are living with ALS to have the courage to talk about their condition and either confirm or refute the findings of this paper to get a clearer picture of what living with ALS looks like across the country.

Future Directions for Investigation

In the future, this study can act as a base for other case studies related to other rare diseases in India. To get a more reliable look at ALS in the country, more case studies, interview and data points will need to be obtained. IAMD could also be another avenue to investigate the efficiency of long-term therapy approaches with ALS patients and see what the effects are for patients at home. The goal of IAMD is for their programs to be helpful for patients looking for an accessible way to conduct therapy at home and learning how to live with their disease, so this would be an appropriate lead to follow for future study. It will also be interesting to look at the implementation of RAN and how financial barriers are effected through the scheme and its application to ALS patients. There is also a large difference between life in rural and urban India and it would be interesting to follow up this study with studies on any differences between rural and urban ALS patients as this was only a survey of Himachal Pradesh ALS patient care. Finally,

based on the observed switch of medical philosophies by patients into Ayurveda, a study looking at ALS patient care in the sphere of Ayurveda could further the knowledge of how these patients are receiving care in India. Overall, this study has opened many new questions about what ALS looks like in India and what it will look like because of institutional change to healthcare in the future of the country.

Political Advocacy on Rare Disease & Community Action

Among developing countries in the world, communicable and non-communicable diseases dominate the narrative on public health priorities. These policies have been influenced by 'do the most good' utilitarianism beliefs across society. While the modern public health model has had great success in treating common disease, it has failed to serve rare disease, especially in India. This desperately needs to change from our findings in this study. As many common communicable and non-communicable diseases have been advocated for by their large-affected-bases, it is because of the extensive medical research that has been put into the development of these efficient treatment regimens that treatment is becoming easier. Simple treatment methods can be brought to communities because they offer widespread benefits across communities. Rare diseases, on the other hand, see only 10% of the population receiving disease-specific treatment (Dharssi 2017). This is mostly due to the little to no financial benefit medical researchers get from researching rare disease and no political incentive in serving these communities. This was observed heavily in the ALS community as questions were asked throughout our surveys by the interviewees about the status of ALS research in the United States and if we carried any recent treatments or findings. These patients wanted was hope in allopathy and modern medicine and the only way that hope can be fulfilled is if we support research and in rare diseases like ALS. Across the world, political interests repeatedly ignore rare diseases due to

only treating small cohorts with rare disease policy. Fundamentally, public health has been built on the World Health Organization's definition as: "the art and science of preventing disease, prolonging life and promoting health through the organized efforts of society." (World Health Organization 2019). With no organized effort from society to advocate for these patients, public health efforts around the world leave ALS patients hopeless and shuffling around hospitals undiagnosed, untreated and dying as was shown in this study. There is a deep need for the field of public health to consider policy approaches to healthcare for rare disease patients and act as the organized effort of society to advocate for rare disease treatment and innovation in medical research.

Healthcare is a social institution incapable of existing within a vacuum untouched by the world of business and personal interests. A community hit especially hard by a lack of business interest is medical research into rare diseases. Researchers will rarely seek treatments or answers to rare diseases out of altruism, which inspired the groundbreaking orphan disease movement in the United States. Pharmaceutical companies following the US Congress' 1983 Orphan Drug Act began to be inspired by tax incentives to research diseases with less than 200,000 sufferers in the country (Khosla 2018). Although great in principle, using this legislation, companies altered their focuses towards rare cancer and hematological diseases to cash in on tax breaks (Khosla 2018). By exploiting tax breaks to research rare cancers, the discoveries and treatments can then be applied to more common cancers to be flipped for immense profit. Rare disease research is not a problem that is only limited in the United States, either. As was reiterated in this study with edaravone in India, pharmaceutical companies are also doing a poor job at making these 'Orphan Drugs' accessible to rare disease patients. One study showed that the average annual cost of an Orphan Drug for a rare disease was $137,782 per patient, clearly limiting rare disease care to

those who can afford it (Khosla 2018). The business of rare disease extends beyond pharmaceuticals and hospitalization is an immense financial burden on patients as well. Studies emerged from Australia and the United States that show rare diseases, while they consisted of 2% and 2.5% of the study cohorts, consisted of 4.6-10.5% and 16-28% of total hospitalization expenditures, respectively (Valdez 2016). This qualitative study of ALS, designated as a rare disease itself, showed that there are financial barriers to receiving pharmaceutical treatments in the case of edaravone in India. With hospitalization costs in many public hospitals being minimal and Indian private hospitals taking inspiration from the American model of hospital care, the financial burden of ALS could be an even greater cost in the private sector. With statistics pointing to the immense financial hurdles rare disease patients are facing, it is important to remind ourselves real people are suffering with rare diseases behind these statistics. These people are forced to work themselves as advocates for their minority communities.

Political interests impede rare disease patients from receiving healthcare. Within the United Nations 2030 Sustainable Development Goals, there was agreement on healthcare treatment to have a fundamental pillar of leaving ‘no one behind in care’ (United Nations 2019). This has been interpreted as the UN’s commitment to treating not only common diseases but also helping patients of rare diseases achieve a quality of life too. Unfortunately, this commitment has been abandoned by India’s public health policies. Despite grassroots patient group advocacy showing great progress towards implementing rare disease policies to help patient groups in the decade, there have been significant setbacks. None have been harder than the failure and abeyance of NPTRD in 2018. This policy, as well as the RAN scheme, should be reinstated, implemented and advertised for ALS patients of low-socioeconomic status as edaravone has a high expense as does having a functional wheelchair, a portable ventilator, and in-home care

services. In-home care services are vital for these patients to have quality of life and that was reiterated by everyone interviewed in Himachal Pradesh associated with care for ALS patients. NPTRD abeyance directly shows the government's focus on only treating the masses in Indian public health by justifying the move through finances. The policy abeyance directly refers to the cost of treating one child with Gaucher's Disease with Enzyme Replacement Therapy compared to being able to treat 400 TB patients (National Policy on the Treatment of Disease 2017). Another move done by the Uttrakhand government was launching a ₹25,00,00,000 project on a search for the *Sanjeevani booti* herb that is believed to be able to cure any neurological illness that was collected by Hanuman and cured Lakshmana in battle. The commitment by the government to fulfilling Ayurveda cures to fulfill political interests in fulfilling religiously backed motives has not been paralleled by government funding into basic research into the etiological features of these rare neurological conditions. With an investment by the Indian government into health at 1.8% of GDP, it is worth asking if investment into policy and research and development can give the 6-8% of the Indian population living with rare disease, access to affordable medicine (National Health Profile 2019). According to Article 21 of the Indian constitution, citizens have been promised the right to health and the government should consider straying away from debates of penny-saving methods of public health and debate a financial investment into life for its citizens (The Indian Constitution 1950).

During a meeting with IAMD in Solan, Himachal Pradesh, we spoke about how investment into rare disease care can help the government and society financially long term with caring for rare disease. They mentioned one of the problems that plague the community is a lack of genetic knowledge about rare diseases. When patients are diagnosed with rare disease, they can be turned away by doctors as had been mentioned previously in cases of ALS because

doctors may not have the time or feel they have adequate resources to provide the care. These people are then going back to their homes and discussing their situation with quacks who then make quick profits from people's vulnerability. The social pressure is immense for these patients and their parents for children diagnosed. The pressure to have a family able-bodied forces many families to continue to have children. They do not realize the root cause of the problem is not existential, yet, it is genetic. Genetic counseling in cases of rare disease may help parents make educated decisions about having more children and save government expenses in rare disease care in the future. Including provisions in future schemes and government policies for many of the non-traditional medical treatments and therapies will be vital in truly helpful for patients.

Findings from this project show the importance of government policy surrounding rare diseases being reinstated, re-evaluated and made in coordination with patients and the states. While palliative care needs definition and national policy in the country, passive euthanasia decisions made by the Supreme Court are not helping this population from achieving quality of life. Findings point towards these patients needing medical devices to improve quality of life. This will require increased investment in health from the national government. In a country that has been plagued with more people dying from a lack of quality of healthcare rather than access to healthcare, improving ALS care will pose a massive challenge for the Indian system as it is incredibly interdisciplinary and unique (Kruk 2018). The government will need to continue to test RAN and ideally, fix the NPTRD. Included in these policies should be provisioned funding and support for patients to have pharmaceuticals, therapy, medical devices, at-home care and genetic counseling for other rare diseases all covered under the provisions of the schemes. Such a comprehensive piece of legislation will be critical to have input from the communities with these rare diseases.

Furthermore, if the government cannot make immediate interventions in the situation, there is a need for awareness of the NGO, Asha Ek Hope's services in rural areas. Suggestions for the NGO's functioning could be to start being able to advise and help educate caretakers with ALS. Our interviews point to caretaker knowledge and ability as being instrumental to quality of life should twenty-four-hour care not to be possible. Also, if the organization could remotely coordinate with patients to deliver these medical devices to them for their usage, it could tremendously help these patients extend their lives living with the disease. As was noted, patients were not being helped by the NGO in rural India which may require communities to intervene in the meantime while the government and NGOs mobilize to help these people.

There were some problems observed in the field that were immediately addressable in the community of Shimla. We have talked with people about establishing a WhatsApp group in Shimla to connect patients and the IGMC and provide caretaker-to-caretaker conversations to help each other with day-to-day caretaking. The caregiver's psychological impact cannot be understated and we created this group with them in mind to maintain their mental health and improve patient quality of care by increasing the caretaker sense of community. One of the caretakers agreed to lead the group and we are hoping for long-term sustainability in the support group as the group grows with more patients going through the IGMC. The group has already contacted the NGO and aims to put these patients who are diagnosed with ALS at IGMC in direct contact with the organization to hopefully receive devices. While this solution hopefully aids in providing patients and caretakers support, there will need to be a greater advocacy effort needed to be taken to municipalities in India.

To combat accessibility issues in Shimla, a conversation was initiated with Municipality Commissioner Shri Pankaj Rai in hopes of making accessibility a high concern and initiate

legislative action as the municipality considers future construction and renovation of buildings in Shimla. Himachal Pradesh already requires public buildings to be accessible, but it was asked that they begin to consider implementing a requirement to consider accessibility in all cases. It was also asked to initiate a program to make the lift program and gondolas free-of-charge for people to be able to access areas such as temples that can offer ALS patients the ability to experience independence again and keep in touch with their spiritual health as well. It is hoped that these changes will be able to make Shimla a more accessible place for the people surveyed, those living there and those visiting Shimla. We hope that these changes can occur to help make the Shimla community a place that is accessible for all and act as a beacon for change in accessibility concerns in cities across the country.

References

1. Brooks, B. et. al. Edaravone in the Treatment of Amyotrophic Lateral Sclerosis: Efficacy and Access to Therapy - A Roundtable Discussion. (2018). *American Journal of Managed Care*, 24. https://www.ajmc.com/journals/supplement/2018/edaravone-treatment-als-access-to-therapy/edaravone-treatment-of-als-efficacy-and-access-to-therapy-roundtable-discussion.
2. Caretaker A (2019, November 18). Personal interview.
3. Choudhury, M. C., & Saberwal, G. (2019). The role of patient organizations in the rare disease ecosystem in India: An interview based study. *Orphanet Journal of Rare Diseases*, *14*(1), 117. https://doi.org/10.1186/s13023-019-1093-6.
4. Dharssi, S., Wong-Rieger, D., Harold, M. (2017). Review of 11 national policies for rare diseases in the context of key patient needs. *Orphanet Journal of Rare Diseases 12*(1)**,** 63. doi:10.1186/s13023-017-0618-0.
5. Gourie-Devi, M., Gupta, R., Sharma, V., Pardasani, V., & Maheshwari, S. (2017). An insight into death wish among patients with amyotrophic lateral sclerosis in India using "Wish-to-Die Questionnaire." *Neurology India*, *65*(1), 46–51. https://doi.org/10.4103/0028-3886.198177.
6. Indian Association of Muscular Dystrophy (IAMD). (2019, December 4). Personal interview.
7. Khosla, D., Patel, F. D., & Sharma, S. C. (2012). Palliative care in India: current progress and future needs. *Indian journal of palliative care*, *18*(3), 149–154. doi:10.4103/0973-1075.105683.
8. Khosla, N., & Valdez, R. (2018). A compilation of national plans, policies and government actions for rare diseases in 23 countries. *Intractable & Rare Diseases Research*, *7*(4), 213–

222. https://doi.org/10.5582/irdr.2018.01085.

9. Kruk, M. E., Gage, A. D., Joseph, N. T., Danaei, G., García-Saisó, S., & Salomon, J. A. (2018). Mortality due to low-quality health systems in the universal health coverage era: A systematic analysis of amenable deaths in 137 countries. *The Lancet*, *392*(10160), 2203–2212. https://doi.org/10.1016/S0140-6736(18)31668-4.
10. Lawyer A. (2019, October 28). Personal interview.
11. Ng, T. S. (2019). Racial differences in experimental pain sensitivity and conditioned pain modulation: A study of Chinese and Indians. *Journal of Pain Research, 12*, 2193-2200. https://doi.org/10.2147/JPR.S197803.
12. *National Health Profile: Central Bureau of Health Intelligence* (No. 10; p. 304). (n.d.). Retrieved from https://cbhidghs.gov.in/index8.php?lang=1&level=1&slid=1068&sublinkid=75&lid=1135&color=1.
13. *NORD IMS Report FNL* (n.d.). Retrieved from https://rarediseases.org/wp-content/uploads/2018/05/NORD-IMS-Report_FNL.pdf.
14. Nurse A. (2019, October 23). Personal interview.
15. Patient A (2019, November 18). Personal interview.
16. Physician A. (2019, October 23). Personal interview.
17. Physician B. (2019, October 23). Personal interview.
18. Physician C. (2019, November 16 & 2019, December 6). Personal interview.
19. Simoens, S. (2011). Pricing and reimbursement of orphan drugs: The need for more transparency. *Orphanet Journal of Rare Diseases*, *6*(1), 42. https://doi.org/10.1186/1750-1172-6-42.

20. Sinha, K. (n.d.). “Nerve disorder affects men more than women” | India News—Times of India. Retrieved November 25, 2019, from https://timesofindia.indiatimes.com/india/Nerve-disorder-affects-men-more-than-women/articleshow/5699295.cms.
21. Social Worker A. (2019, October 23). Personal interview.
22. Sondhi, S., Sharma, S., Kaushal, S. S., Mehta, A., & Banayal, V. (2018). The Profile of Amyotrophic Lateral Sclerosis in Natives of Western Himalayas: Hospital-Based Cohort Study. *Journal of neurosciences in rural practice*, *9*(3), 305–311. doi:10.4103/jnrp.jnrp_8_18.
23. The Constitution of India, 26 January 1950, available at: https://www.refworld.org/docid/3ae6b5e20.html [accessed 4 November 2019].
24. Thomas, Priya & Sadasivan, Arun & Balasubramanium, Bhuvaneshwari & Preethish-Kumar, Veeramani & Nashi, Saraswati & Polavarapu, Kiran & Krishna, Gopalkrishna & Vengalil, Seena & Rajaram, Prakashi & Nalini, Atchayaram. (2018). Caregiver burden and quality of life of patients with amyotrophic lateral sclerosis in India. *Amyotrophic Lateral Sclerosis and Frontotemporal Degeneration*. *19*. 1-5. doi:10.1080/21678421.2018.1482353.
25. United Nations Sustainable Development. (n.d.). Retrieved November 5, 2019, from https://www.un.org/sustainabledevelopment/sustainable-development-goals/.
26. Valdez, R., Ouyang, L., & Bolen, J. (2016). Public Health and Rare Diseases: Oxymoron No More. *Preventing Chronic Disease*, *13*. https://doi.org/10.5888/pcd13.150491.
27. World Health Organization: Public Health. (2019, November 3). Retrieved November 4, 2019, from World Health Organization website: http://www.euro.who.int/en/health-topics/Health-systems/public-health-services.
28. Yadav C, Chaubey S, Kurele R, Semwal DK (2017). Sanjeevani booti - A majestic and

elusive all curing divine herb in epic Ramayana. *Journal of Conventional Knowledge and Holistic Health*, 1 (1), Article ID 173.