The Intense Psychological Burden of ALS, the Enduring Strength of People Living With ALS, and the Tools We Can Use to Help

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We are making progress in understanding the biology of amyotrophic lateral sclerosis (ALS) at an increasingly rapid pace. An unprecedented number of trials in ALS are currently underway. Drugs entering clinical development now are far more likely to succeed than in the past, as evidenced by the fourth FDA approval of an ALS disease-modifying therapy. These advances have lent a remarkable optimism to the field, yet, at present, we do not have a cure.

The emotional toll the disease takes on a person living with ALS (pwALS) may be severe, starting from the time of symptom onset and continuing through diagnosis and into the final stages of the disease. Although multidisciplinary ALS clinics may help to provide support, to date there are no ALS-specific psychological interventions that can be offered to pwALS.

In this issue of JCP, Lester and colleagues[1] wrestle with these issues and their experience treating "Michael," a pwALS diagnosed 12 years prior, for recurrent major depression and adjustment challenges related to his ALS in the setting of an ongoing divorce. The authors describe their psychotherapeutic and psychopharmacologic work with this patient, his gains and setbacks, and the impact on themselves as providers—raising essential questions about the emotional toll of pwALS and how providers can work together to improve care of pwALS. The authors raise many salient questions requiring resolution. What are the specific emotional impacts of diagnosis and disease progression? How do advances in ALS treatment and in our understanding of genetics impact pwALS and asymptomatic gene carriers and their emotional journeys? Most importantly, how can we, as care teams, work together to support pwALS and their families?

Diagnosis

A diagnosis of ALS throws open a set of concerns heretofore never considered by a pwALS. Receiving a diagnosis is incredibly emotional—whether it is a new diagnosis, which can be entirely unexpected, or a diagnostic confirmation, which can extinguish the final ray of hope for an alternate and curable diagnosis. Delays in diagnosis can also impact the response when the diagnosis is finally given.

As we have more therapies for ALS, know more about lifestyle changes required to slow progression, and better understand the genetics of ALS, the diagnostic visit becomes ever more laden with critical, time-sensitive information and choices, heightening the emotional stakes.

PwALS frequently recount their experience in diagnostic visits as an emotionally traumatic episode. They often recount a deep focus on small aspects of the environment, a relative lack of organized memories about the discussion, and sometimes a sense of depersonalization occurring at or following the visit. PwALS may also remember experiencing a tremendous amount of fear,[2] triggering a near fight-or-flight response, leading to high levels of anxiety, depression, or avoidance.

Without support, this time is overwhelming. Depression rate and suicide risk may be higher earlier in the disease and closer to the time of diagnosis.[3–5] Our clinical observations have also led us to recognize diagnosis as a time of tremendous emotional stress, when internal and external supports either strengthen or disintegrate and information often cannot be absorbed. Currently, we do not know what the optimal support is, due to a lack of focused studies on the emotional journey of pwALS.[6] To fill the need for support, our ALS clinic has implemented a visit with our ALS nurse practitioner or physician in the near weeks following diagnosis to review the diagnosis and the treatment options and to support emotional distress. We also focus on implementation of best practices in breaking a diagnosis like ALS and involvement of palliative care neurologists to help provide information and supportive resources that center of discussions on quality of life.[7]

Genetics

Adding the uncertainty of genetic testing to the diagnosis of ALS may increase the anxiety and depression that can surround the diagnosis. Anxiety, and in some cases guilt, may surround identification of an ALS-causative mutation because of the potential of passing a mutation to subsequent generations. Furthermore,

as we identify more cases of people with Mendelian genetic forms of ALS through standard testing, we also identify a larger cohort of asymptomatic people who carry ALS causative genes. This has opened an important new population for monitoring and research and is also creating a growing population of individuals facing the psychological implications of discovering they carry a gene that could cause ALS. Specific psychological counseling programs for these individuals have yet to be established, although ongoing studies are working to define appropriate psychological supports (eg, the Dominant Inherited ALS Network and the ALS Families and Pre-symptomatic Familial ALS projects).

Disease Progression

ALS is a relentlessly progressive disease, but there are unpredictable "plateaus" during which functional loss is minimal and "cliffs" during which functional loss is rapid. The disease progression is predictable in the general sense and unpredictable in the specific sense. PwALS and caregivers are left constantly adjusting their lifestyle to ever-increasing loss of physical function. They frequently tell us that, with effort, they could adjust to their current physical limitation, but that the constant loss of more physical capacity can be emotionally difficult to account for.

Because the physical progression of the disease is highly variable between pwALS, and because the psychological resilience and supports vary widely between pwALS, it can be hard to map out a stereotyped psychological support and treatment program for pwALS broadly. Furthermore, because ALS can cause bulbar muscle weakness and resultant dysarthria, psychotherapy can be challenging and require modifications to be successful. Familial and social support structures are highly variable and evolve as disease progresses and caregiver burden increases; pwALS who lack support and are more isolated may be at higher risk for depression and suicide, although the literature is not conclusive.[3] Loss of autonomy and development of bulbar symptoms may contribute to depression at later stages of the illness.[4]

Rates of depression may be higher in the ALS population over the course of the disease,[4] but symptoms of depression may also overlap with progressive ALS symptoms, leading to underrecognition of depression.[8] Not surprisingly, more experienced ALS providers may be able to more accurately to estimate quality of life and depressiveness than less experienced counterparts.[8] Evaluation of mood can also be complicated by the presence of pseudobulbar affect, which causes uncontrolled crying, even without underlying depression.[9] Fatigue, one of the most common symptoms of ALS,[10] can also mimic some symptoms of depression. The risk of suicide in pwALS is higher than the general population, perhaps 6-fold as in certain studies.[3]

In our experience, for pwALS without preexisting anxiety or depression, psychopharmacology for new symptoms can be quite successful, even with low doses of SSRIs or SNRIs. For those with a history of major depressive disorder, generalized anxiety, or other mental illness, collaboration between ALS providers and psychopharmacologists is helpful and can be a model for more widespread collaboration.

Path Forward

Having identified some of the significant emotional and psychiatric challenges facing pwALS, it is clear that well-designed and individualized psychiatric and psychological support programs could benefit pwALS. The challenge now is to develop models for therapy that respect the physical limitations imposed by ALS, the psychological needs of pwALS and their caregivers, and the psychopharmacologic tools that are safest and most effective for pwALS. Ongoing communication between ALS-treating providers and psychiatrists and psychologists will help tailor the therapies to each person and respond to the evolving nature of the disease.[6,11]

Limited resources are a consideration in developing a successful program. The costs of multidisciplinary clinics are high. Many psychiatric therapies such as cognitive behavioral therapy are low in reimbursement. Mental health providers are in short supply. PwALS and their families are often under great financial and time pressures. The implementation of telehealth may help. In some cases, genetic counselors will also play a critical role in these collaborations. Earlier psychological screening and identification of preexisting psychiatric symptoms or illness may help identify pwALS at highest risk for mental health concerns. Screening could consist of clinical conversations or ALS specific surveys of depression and anxiety to help prompt early referrals. Finally, awareness is key. Creating materials to educate mental health providers about ALS could help prepare them for the challenges and opportunities ahead. Case reports like this one can increase awareness and understanding of the issue, and more systematic testing of proactive models of therapy will also help to improve care.

Drs Lester and Vitolo poignantly highlight that the disease also impacts those of us who care for, and about, pwALS. As providers, we are welcomed into an individual's most private conversations and bear witness to the greatest challenges a person will ever face, offering what help we can, while knowing we would like to do much more. As ALS clinical researchers, we see scientific advances that are coming and can participate in the quest to overcome ALS, which buoys our spirits. Perhaps equally important, as ALS clinicians, we see a beauty and strength of human spirit in the face of great adversity that few others ever see. The question, becomes not, "How can we do this job?" but rather, "How could we not?"

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