

Introduction

Motor neurone disease (MND), also referred to as amyotrophic lateral sclerosis (ALS) and Lou Gehrig disease in the US, is a life-limiting neurodegenerative condition. The condition progressively destroys the motor neurons in the brain and spinal cord and alters an individual's ability to control voluntarily their muscle movements, leading to paralysis, swallowing difficulties, respiratory failure and, ultimately, death (King, Mulligan & Stansfield, 2014). The effects of MND are not limited to motor functions; behavioural difficulties, cognitive impairment (McCluskey et al., 2009; Strong et al., 1999), emotional difficulties (depression, anxiety and anger) and involuntary changes in mood (Orrell, 2016) are also common. With a prevalence rate for North America and Europe of around two per 100,000 of the population (Worms, 2001), median survival rates following symptom onset are generally only two to four years, with only 10%-20% of individuals surviving past 10 years (Chiò et al., 2009).

Current interventions for individuals with MND mainly focus upon the physical aspects of the condition, to maintain physical and biological functioning and quality of life for as long as possible (Andersen et al., 2012). However, a recent mixed-method review on the supportive needs of this group (Oh & Kim, 2017) reported that of the 37 studies included, only around half discussed psychosocial needs, concluding a significant need for more psychological, social and emotional support, alongside physical and practical assistance. The authors also proposed that the psychological impact of receiving, adjusting and coping with this diagnosis should be considered equally with the physical impact of the condition.

Individuals with a diagnosis of MND are reported to experience heightened psychological distress and decreased well-being (Hogg, Goldstein & Leigh, 1994; Lou, Reeves, Benice & Sexton, 2003; Montgomery & Erikson, 1987; Tedman, Young & Williams, 1997; Vignola et al., 2008). Although several factors undoubtedly contribute to this (e.g.,

biological, social, psychological and ‘spiritual’ suffering: Ganzini, Johnston & Hoffman, 1999; social withdrawal: Rigby et al., 1999; and physical impairment: Hunter, Robinson & Neilson, 1993; Hogg et al., 1994), recent research in other physical health conditions has suggested that stigma may be an important variable to consider. This has been identified as important in health conditions with perceivable physical effects such as epilepsy (Baker, Eccles & Caswell, 2018), Parkinson’s disease (Simpson, MacMillan & Reeve, 2012) and multiple sclerosis (Broersma, Oeseburg, Dijkstra & Wynia, 2018).

The term stigma was originally defined by Goffman (1963) as “an undesired differentness” (p. 5) and this has since been developed to include two concepts of stigma: felt and enacted (Scambler & Hopkins, 1986). ‘Felt stigma’ refers to a feeling of shame about being different and feeling that discrimination for this difference will occur, whereas ‘enacted stigma’ refers to actual experience of this discrimination. Link and Phelan (2001) further developed the concept and described stigma as a set of components which include labelling, stereotyping, separation, status loss, and discrimination. Therefore, the term stigma can encompass a range of negative actions and associations that can be attributed to an individual based on their perceived differentness. In relation to MND, only one study has quantitatively assessed stigma (van der Beek, Bos, Middel & Wynia, 2013). Although the authors found that stigma was a major predictor of poorer quality of life, with ‘felt stigma’ a stronger predictor than ‘enacted stigma’, the study’s outcome measure was quality of life, rather than psychological distress in particular, and individuals diagnosed with MND only made up 9% of the total number of participants. Furthermore, no research has been conducted to assess the role that felt stigma plays in mediating the relationship between enacted stigma and psychological distress, as theorised in the self stigma model proposed by Corrigan and colleagues (Corrigan & Watson, 2002; Corrigan, Watson & Barr, 2006).

Another variable which has been less studied in motor neurone disease as opposed to other neurodegenerative conditions is social support. Research has identified social support as a significant predictor of psychological distress (conceptualised as depression and quality of life; Matuz, Birbaumer, Hautzinger & Kübler, 2010). Furthermore, longitudinal research has also identified social support as a significant predictor of depression and quality of life (Matuz, Birbaumer, Hautzinger, & Kübler, 2015) and depression and anxiety (Goldstein, Atkins, Landau, Brown and Leigh, 2006). Despite research identifying social support as a significant predictor of psychological distress for individuals with MND, no research has been conducted, to the best of the authors' knowledge, to assess whether social support acts as a moderator between MND related stress and psychological distress. This moderating effect of social support would suggest that social support could act as a 'buffer' on the relationship between MND related stress and psychological distress, as proposed in the stress buffering model (Barrera, 1986; Cohen & Wills, 1985; Cutrona & Russell, 1990; Thoits, 1986).

Consequently, this study aims to investigate the mechanisms through which perceived stigma and social support influence the relationship between MND related stress and psychological distress for people with MND. The research aim was to investigate whether increased levels of perceived stigma and lower levels of social support influenced the relationship between MND related stress and psychological distress. It was hypothesised that both enacted and felt stigma would be significant predictors of psychological distress in individuals with MND, and that felt stigma would significantly mediate the relationship between enacted stigma and psychological distress (depression, anxiety and stress). Furthermore, it was hypothesised that social support would significantly moderate the relationship between MND related stress and psychological distress.

Method

Design

A quantitative cross-sectional survey design was used to investigate: a) whether felt stigma mediated the relationship between enacted stigma and psychological distress; and b) if social support had a moderating effect on the relationship between MND related stress (conceptualised as scores on a measure of both physical functioning and enacted stigma) and psychological distress (depression, anxiety and stress) in individuals with a diagnosis of MND. Both the mediation and moderation analyses were conducted using Hayes' PROCESS Tool (Hayes, 2018).

Participants

Individuals aged 18 years or over who had a diagnosis of MND/ ALS/ Lou Gehrig disease and who could complete an online survey written in English (either alone or with assistance from another person) were eligible to take part. An opportunistic sampling method was employed as participants volunteered to participate following advertisement of the study details online, using social media and through international organisations. If a participant wished to take part in the study, then they accessed the study link provided in the study advert and were given the opportunity to read and download the participant information sheet. Eligibility for the study was based on self-report and was recorded through a demographic questionnaire at the beginning of the survey.

The literature was consulted to determine the required sample size to detect the mediated effect and based on using an indirect effect method of mediation with bias-corrected bootstrapping and medium effect size in both arms ($a=.39$ and $b=.39$), 71 participants were needed for 80% power (Fritz & MacKinnon, 2007). For moderation analyses, effect sizes are typically small (Aguinis, Beaty, Boik, & Pierce, 2005), with suggested sizes for f^2 of 0.005, 0.01, and 0.025 for small, medium, and large effects,

respectively (Kenny, 2018). A large effect size would need 316 participants to detect an effect (G^* power; Faule, Erdfelder, Buchner & Lang, 2009). A total of 77 participants were recruited; 34 women and 43 men (M age = 59.14). Of the 94 participants who opened the survey, 84 completed the demographic data, with only 80 of these continuing to complete the outcome measures. Of these, only 78 completed all the measures (two participants did not complete the physical functioning measure and their data were withdrawn). A further participant's data were withdrawn due to not meeting the inclusion criteria of being completed by an individual with a diagnosis of MND. See Table 1 for participants' self-reported demographic characteristics.

Insert Table 1 here please

Procedure

Ethical approval was gained from the first author's host academic institution after input from service users. Recruitment took place online from a variety of sources, mainly through international organisations supporting people with MND. This was achieved by sharing the study advert and link on social media (Facebook and Twitter) pages by the Motor Neurone Disease Association (MNDA) Australia, MNDA UK, MNDA New Zealand, MNDA Scotland, ALS Association Canada, ALS Society Quebec, Minds and Movement, and the first author's own contacts. An online survey was constructed using Qualtrics software which was used to collect the data. Prior to completing the survey each participant accessed the participant information sheet and then consented to take part. Following completion of the questionnaires, a debrief sheet was available which also provided the

contact details of organisations who could provide support if participation caused any distress.

Measures

To control for potential confounders, measures were included to assess demographic variables (age and gender), along with the three variables of interest (physical functioning, social support and stigma). One measure was used to assess the three outcomes of psychological distress (measuring depression, anxiety and stress). To situate the sample additional variables were collected including nationality, relationship status, time since symptom onset and time since diagnosis.

Physical Functioning

The Self-Administered Amyotrophic Lateral Sclerosis Functional Rating Scale Revised (SA-ALSFRS-R; Cedarbaum et al., 1999; Montes et al., 2006) includes 12 questions which assess the domains of motor function, bulbar symptoms and breathing ability in individuals with MND. Individual items are rated on a scale of 0 to 4 with a total score range of 0 to 48; higher scores indicate higher levels of physical functioning. Although there is no current evidence regarding the reliability and validity of the SA-ALSFRS-R, there is evidence regarding the clinician administered ALSFRS-R. This has been shown to be a reliable and valid measure by the authors during development (Cedarbaum et al., 1999). Montes et al. (2006) compared the use of the SA-ALSFRS-R to the clinician administered ALSFRS-R and reported an intraclass correlation coefficient score of $r = 0.93$, implying that the self-administered version is as reliable as the clinician administered version. This measure was chosen as it is aimed specifically at individuals with a diagnosis of MND to assess their physical functioning and symptom severity. The use of the ALSFRS-R instrument online compared to on-site face-to-face assessment was assessed by Maier et al.

(2012) and their results supported the use of the measure online, due to a highly significant correlation between on-site evaluation and online testing ($r = 0.96$).

Social Support

The Multidimensional Scale of Perceived Social Support (MSPSS; Zimet, Dahlem, Zimet & Farley, 1988) is a 12-item measure scored on a scale from 1 (very strongly disagree) to 7 (very strongly agree), with a total score range of 12 to 84; the scale contains three subscales (four items in each): significant other, family and friends (scores range from 4 to 28 for each subscale). Both the total scale and for the individual subscales can be used, with higher scores indicating higher levels of social support. The authors report highly acceptable Cronbach's coefficient alpha co-efficients and good test-retest reliability. The scale is relatively short, easy to complete and measures individuals' levels of social support from different sources.

Stigma

The Stigma Scale for Chronic Illness (SSCI, Rao et al., 2009) comprises 24 questions with two subscales; one scale for felt stigma (labelled self-stigma; 13 questions) and one scale for enacted stigma (11 questions). Each item is scored on a scale of 0 (never) to 4 (always) with a total score range of 0 – 96 for the full scale, 0 – 52 for the felt scale and 0 – 44 for the enacted scale. Higher scores indicate a higher level of stigmatisation. Internal reliability for this scale was assessed by the authors and was found to be highly acceptable. The measure was specifically developed for people with chronic neurological illnesses, including individuals with MND. The total score was used alongside the two subscales (felt and enacted stigma) for the correlational analyses for this study and the two subscales were used in the regression analyses of this study. As part of the validation process, it was administered online (Rao et al., 2009).

Psychological Distress

The Depression, Anxiety and Stress Scale (DASS-21; Lovibond & Lovibond, 1995) is a 21-question scale with three subscales, depression, anxiety and stress (seven questions for each subscale). These are scored on a range of 0 (never) to 3 (almost always) with a total range of 0 – 21 for each of the subscales; higher scores indicate a higher level of distress and cut-offs have been created to categorise scores as: “normal”, “mild”, “moderate”, “severe” or “extremely severe”. The scale has excellent internal reliability both in its full form and in the three subscales (Antony, Bieling, Cox, Enns, Swinson & Haynes, 1998; Henry & Crawford, 2005). The measure has been used online in previous research with individuals with MND (Lillo, Mioshi, Zoing, Kiernan & Hodges, 2011; Caga, Ramsey, Hogden, Mioshi & Kiernan, 2015).

Data Analysis

Data analysis was completed using SPSS (Version 26) and Hayes PROCESS macro Version 3.4.1. The sample was checked for missing data prior to any analysis being conducted; seven participants had not provided a response to the same question on the SSCI: “people with my illness lost their jobs when their employers found out”. This missing datum was replaced with the mean value of this specific subscale of the SSCI (enacted stigma subscale) for each person. No other missing data was identified in the sample. Outliers were identified using boxplots and scores were checked for errors. Given the relatively small sample size, a less conservative method (dividing the skewness or kurtosis value by its standard error) was used to assess for skewness and kurtosis (Field, 2005). Using parameters of -3 to +3, the only scale not normally distributed was the MSPSS total score.

As the MSPSS total score was not normally distributed, non-parametric correlations were calculated using Spearman’s correlation coefficients to identify relationships.

Mediation analyses were then conducted; in each analysis the predictor variable was enacted stigma, the mediator variable was felt stigma and the outcome variable was either depression, anxiety, or stress. Each analysis was based on 5000 bootstrap samples to estimate the confidence intervals. Moderation analyses were then conducted to assess the moderating effects of social support on the relationship between MND related stress and psychological distress. In each analysis the predictor variable to represent MND related stress was either enacted stigma or physical functioning and the outcome variable was either depression, anxiety or stress, with social support as the moderating variable.

All regressions analyses which contributed to the mediations and moderations were checked to ensure that they met assumptions of independent errors, homoscedasticity and normalised residuals and that no multicollinearity was present.

Results

The mean scores, standard deviations and Cronbach's alpha scores for each measure are reported in Table 2.

Insert Table 2 here please

The mean (*SD*) score for the SA-ALSFRS-R was 29 (8.62), indicating that levels of independent functioning were within the mid-point range. The majority of scores fell within the 'mild to moderate' category (43%, $n = 33$), with 'moderate to severe' (30%, $n = 23$), 'advanced disease' (16%, $n = 12$) and the 'minimal to mild' category (12%, $n = 9$).

The mean score for the MSPSS total ($M = 66.47$; $SD = 15.66$) indicated that levels of social support were towards the higher end of the scale for this sample (possible range: 12-

84). Moreover the mean score for the SSCI total score ($M = 32.83$; $SD = 16.12$) indicated that levels of stigma were quite low in this sample (possible range: 0 - 96, with higher scores indicating higher levels of stigma).

Regarding the DASS scores, for the depression subscale, the majority of scores were in the 'normal' category (70%, $n = 54$), followed by the 'mild' (18%, $n = 14$), and the 'moderate' category (12%, $n = 9$). For the anxiety subscale, all the scores fell within the 'normal' category (100%, $n = 77$). For the stress subscale, the majority of scores were in the 'normal' category (62%, $n = 48$), followed by 'moderate' (22%, $n = 17$), 'mild' (9%, $n = 7$), 'severe' (5%, $n = 4$) and finally the 'extremely severe' category (1% $n = 1$). This indicates that the majority of the sample had non-clinical levels of stress, anxiety and depression.

Correlations

Spearman's r_s correlation coefficients were calculated to determine the relationships between the variables; see Table 3.

Insert Table 3 here please

As can be seen in Table 3, none of the demographic or clinical variables correlated significantly with the three outcome variables (depression, anxiety or stress). However, statistically significant relationships were found between the two stigma subscales and social support variables and all three outcome variables. Associations were in the predicted direction with social support (total score) negatively correlated with depression, anxiety and stress and stigma (total score and the two subscales) positively correlated with the three outcome measures.

Mediation Analyses

As physical functioning significantly correlated with both stigma variables, this was initially included in the mediation models as a covariate. Upon further inspection, this variable did not make any significant contribution to the model or impact the findings greatly, therefore, this was not included in the final models. In each analysis the predictor variable was enacted stigma, the mediator variable was felt stigma and the outcome variable was either depression, anxiety, or stress. Mediation analyses were conducted to assess the mediating effects of felt stigma; see Table 4 and Figures 1 to 3 for the main findings of these analyses.

Insert Table 4 here please

Insert Figure 1 here please

Insert Figure 2 here please

Insert Figure 3 here please

Mediation model for depression.

The total effect of the model was significant ($c = .401$, 95% CI [.259, .543], $p < .001$). Higher enacted stigma predicted higher felt stigma ($a = .941$, 95% CI [.683, 1.198], $p < .001$) and higher felt stigma predicted higher depression scores ($b = .316$, 95% CI [.211, .420], $p < .001$). A significant indirect effect was found for enacted stigma through felt stigma on depression scores ($ab = .297$, 95% CI [.191, .429]). The effect size of the completely standardised indirect effect was .404. The direct effect of the model did not remain significant when controlling for the effect of the mediational variable of felt stigma ($c' = .104$, 95% CI [-.049, .257], $p > .05$).

Mediation model for anxiety.

The total effect of the model was significant ($c = .196$, 95% CI [.080, .312], $p < 0.01$). Higher enacted stigma predicted higher felt stigma ($a = .941$, 95% CI [.683, 1.198], $p < .001$) and higher felt stigma predicted higher anxiety scores ($b = .206$, 95% CI [.113, .300], $p < .001$). A significant indirect effect was found for enacted stigma through felt stigma on anxiety scores ($ab = .1941$, 95% CI [.092, .313]). The effect size of the completely standardised indirect effect was .358. The direct effect of the model did not remain significant when controlling for the effect of the mediational variable of felt stigma ($c' = .002$, 95% CI [-.134, .138], $p > .05$).

Mediation model for stress.

The total effect of the model was significant ($c = .365$, 95% CI [.242, .488], $p < .001$). Higher enacted stigma predicted higher felt stigma ($a = .941$, 95% CI [.683, 1.198], $p < .001$) and higher felt stigma predicted higher stress scores ($b = .193$, 95% CI [.092, .294], $p < .001$). A significant indirect effect was found for enacted stigma through felt stigma on stress scores ($ab = .182$, 95% CI [.069, .278]). The effect size of the completely standardised indirect

effect was .281. The direct effect of the model remained significant when controlling for the effect of the mediational variable of felt stigma ($c' = .184$, 95% CI [.036, .332], $p < .05$).

Moderation Analysis

To assess the moderating effects of social support on the relationship between MND related stress and psychological distress, moderation analyses were conducted. In each analysis the predictor variable to represent MND related stress was either enacted stigma or physical functioning and the outcome variable was either depression, anxiety, or stress with social support as the moderating variable. There was no significant moderation effect of social support found in any of the models. For MND related stress, conceptualised as either enacted stigma or physical functioning, neither of the models including stress were significant ($p = .179$ for enacted stigma; $p = .237$ for physical functioning), neither of the models including anxiety were significant ($p = .789$; $p = .816$ respectively) and nor were those models including depression ($p = .164$; $p = .379$ respectively).

Discussion

The present study investigated the impact of social support and perceived stigma on the relationship between MND related stress and psychological distress in individuals with MND. Statistically significant correlations were found between social support, stigma (total score, self (felt) subscale and enacted subscale) and all the outcome variables in the predicted directions. The correlational findings suggest that individuals with higher levels of social support had lower levels of psychological distress (depression, anxiety and stress) and that individuals with higher levels of stigma (both felt and enacted) also experienced higher levels of psychological distress.

To explore these relationships further, mediation and moderation analyses were conducted. In relation to the role of felt stigma in mediating the relationship between MND related stress (enacted stigma) and psychological distress (depression, anxiety and stress),

analyses yielded significant results. The effect of felt stigma as a mediator of the relationship between enacted stigma and psychological distress was significant. Each of the mediation models found a significant indirect effect via this mediator. This suggests that the relationship between enacted stigma and psychological distress is mediated by the individual's levels of felt stigma. These findings are consistent with the theoretical model of self stigma proposed by Corrigan and colleagues (Corrigan & Watson, 2002; Corrigan, Watson & Barr, 2006) whereby public attitudes (enacted stigma) produce personal responses and self stigmatisation which then leads to negative consequences for the individual.

This process begins when an individual with a condition that induces stigmatisation experiences discrimination (enacted stigma) which makes them aware of the negative stereotypes attached to their condition. This awareness of the negative stereotype is conceptualised as felt (perceived) stigma and the actual experience of discrimination (e.g. social exclusion) is conceptualised as enacted stigma. The process can lead to the individual agreeing with the negative stereotypes and then internalising the stereotype. This internalisation is conceptualised as self stigma, which then results in consequences for the self (such as psychological distress). The results of the present study lend support to this model as it identified a significant indirect effect of felt stigma on the relationship between enacted stigma and depression, anxiety and stress. However, the direct path between enacted stigma and stress also remained significant after mediation analysis, suggesting that the actual experience of discrimination also relates to stress levels for individuals with MND independent of the process of internalisation of the negative stereotype. It is interesting to note that theoretical accounts of the effects of disablism from other disciplines, such as the social model of disability (Thomas, 2007) and the later concept of psycho-emotional disablism (see Simpson & Thomas, 2014), would also predict a direct route from hostile disablist attitudes to well-being and an indirect one via the internalisation of these attitudes.

Moderation analyses revealed no significant effect for the role of social support as a moderator on the relationship between MND related stress (enacted stigma or physical functioning) and psychological distress (depression, anxiety and stress).

Limitations

While this study has used established procedures to measure mediation, it is still acknowledged that this approach is limited in confirming causal inferences (Levin, 2006). Sample size is a further limitation in this study, as only a relatively small sample size was achieved. The suggested sample size to achieve a medium effect for the moderation analysis was 472 participants, and this study only recruited 77 participants, falling well below the suggested requirement. Due to the relative rarity of the condition, with prevalence rates for North America and Europe of around two per 100,000 of the population (Worms, 2001), and its rapid degenerative effect, recruiting enough participants to achieve a sample size to test moderation was challenging. While an online survey method was used to extend recruitment internationally, this did rely on the cooperation of organisations and support services. For example, only one local organisation in the US shared this information, resulting in only a small proportion (3%) of participants from this country.

Levels of stigma were quite low in this sample and most of the sample had 'normal' levels of stress, anxiety and depression. A review of psychosocial aspects of MND (McLeod & Clarke, 2007) identified varying levels of depression and anxiety within MND populations; these ranged from 0% to 50% for depression (defined as moderate to severe) and 11% to 26% for anxiety. Therefore, given the relatively high variability, it is unclear whether the current sample may be considered representative although the present findings may not be applicable to samples with higher levels of stigma and psychological distress. Furthermore, participants in this study were mainly longer surviving individuals, which again might limit

generalisability, with the very low levels of depression, anxiety and stress not similar in more recently diagnosed individuals.

Finally, this study had a very highly selected sample as, despite at least 16 countries included, only a small number from each country answered the questionnaire. Given the online format, this also restricted or privileged those individuals interested in this topic and who had access to an online platform.

Clinical Implications

The findings from this study provide implications for clinical practice. In particular, results suggest that if psychological distress is identified in an individual with MND, then it may be beneficial to use both social and psychological factors to inform formulations and be aware of the potential of both routes to impact when considering a comprehensive intervention plan.

Interventions aimed at targeting stigma often operate on several levels: intrapersonal, interpersonal, organisational/ institutional, community and governmental/ structural (Heijnders & Van Der Meij, 2006). Systematic reviews have identified that the most effective interventions are aimed at the intrapersonal, interpersonal and community levels (Heijnders & Van Der Meij, 2006; Rao et al., 2019). Effective intervention strategies for reducing stigma for conditions such as HIV, mental health diagnoses and leprosy include education (e.g., Ngoc, Weiss & Trung, 2016), counselling (e.g., Lusli et al., 2016), cognitive behavioural therapy (Corrigan & Calabrese, 2005), social marketing (Henderson et al., 2012), drama therapy (Orkibi, Bar & Eliakim, 2014) and social support groups (Thurman, Jarabi & Rice, 2012), and combinations of these (e.g., Uys et al., 2009). Moreover, research suggests that mindfulness is positively associated with stigma resistance in individuals with a psychiatric diagnosis (Chan, Lee & Mak, 2018) suggesting that mindfulness-based interventions may be beneficial in bolstering the self against the effects of enacted stigma.

Mindfulness-based interventions have also been identified as effective in reducing psychological distress for individuals with a diagnosis of MND (Pagnini et al., 2015; Pagnini et al., 2017) which could make them useful for a number of adaptive purposes.

However, individually focused interventions are clearly not sufficient on their own to tackle stigma and, for example, using the concept of psycho-emotional disablism (Thomas, 2007), it is also important to look at addressing the limitations and barriers society imposes on individuals with impairments. Socially engaged activism, while not the usual intervention route for health and clinical psychologists (Simpson & Thomas, 2014), is important in effecting societal change to highlight and break down these structural barriers.

Conclusion

Mediation analyses identified significant findings for the indirect effect of felt stigma on the relationship between MND related enacted stigma and psychological distress (depression, anxiety and stress), while a direct route from enacted stigma to psychological distress also remained in the case of stress (one of the three relationships examined). Moderation analysis revealed no significant role for social support as a moderator of the relationship between MND related stress and psychological distress. These findings should be used to improve interventions for individuals with a diagnosis of MND as they highlight the importance of considering both social and psychological factors when psychological distress has been identified. For individuals with MND, the roots and causes of psychological distress are complex – and our approach to intervention needs to acknowledge this.

References

- Aguinis, H., Beaty, J. C., Boik, R. J., & Pierce, C. A. (2005). Effect Size and Power in Assessing Moderating Effects of Categorical Variables Using Multiple Regression: A 30-Year Review. *Journal of Applied Psychology, 90*(1), 94-107. doi:10.1037/0021-9010.90.1.94
- Andersen, P. M., Abrahams, S., Borasio, G. D., de Carvalho, M., Chiò, A., ... & Petri, S. (2012). EFNS guidelines on the clinical management of amyotrophic lateral sclerosis (MALS)—revised report of an EFNS task force. *European Journal of Neurology, 19*(3), 360-375.
- Antony, M. M., Bieling, P. J., Cox, B. J., Enns, M. W., & Swinson, R. P. (1998). Psychometric properties of the 42-item and 21-item versions of the Depression Anxiety Stress Scales in clinical groups and a community sample. *Psychological Assessment, 10*(2), 176.
- Baker, D., Eccles, F. J., & Caswell, H. L. (2018). Correlates of stigma in adults with epilepsy: a systematic review of quantitative studies. *Epilepsy & Behavior, 83*, 67-80.
- Barrera Jr, M. (1986). Distinctions between social support concepts, measures, and models. *American journal of community psychology, 14*(4), 413-445.
- Broersma, F., Oeseburg, B., Dijkstra, J., & Wynia, K. (2018). The impact of self-perceived limitations, stigma and sense of coherence on quality of life in multiple sclerosis patients: results of a cross-sectional study. *Clinical rehabilitation, 32*(4), 536-545.
- Caga, J., Ramsey, E., Hogden, A., Mioshi, E., & Kiernan, M. C. (2015). A longer diagnostic interval is a risk for depression in amyotrophic lateral sclerosis. *Palliative & Supportive Care, 13*(4), 1019-1024.

- Cedarbaum, J. M., Stambler, N., Malta, E., Fuller, C., Hilt, D., Thurmond, B., ... (1999). The ALSFRS-R: a revised ALS functional rating scale that incorporates assessments of respiratory function. *Journal of the neurological sciences*, *169*(1-2), 13-21.
- Chan, K. K. S., Lee, C. W., & Mak, W. W. (2018). Mindfulness model of stigma resistance among individuals with psychiatric disorders. *Mindfulness*, *9*(5), 1433-1442.
- Chi, P., Li, X., Zhao, J., & Zhao, G. (2014). Vicious circle of perceived stigma, enacted stigma and depressive symptoms among children affected by HIV/AIDS in China. *AIDS and Behavior*, *18*(6), 1054-1062.
- Chiò, A., Logroscino, G., Hardiman, O., Swingler, R., Mitchell, D., Beghi, E., ... & Eurals Consortium. (2009). Prognostic factors in ALS: a critical review. *Amyotrophic Lateral Sclerosis*, *10*(5-6), 310-323.
- Cohen, S., & Wills, T. A. (1985). Stress, social support, and the buffering hypothesis. *Psychological bulletin*, *98*(2), 310.
- Corrigan, P. W., Watson, A. C. (2002). The paradox of self-stigma and mental illness. *Clinical Psychology: Science and Practice*, *9*(1), 35-53.
- Corrigan, P., Watson, A., Barr, L. (2006). The self-stigma of mental illness: Implications for self esteem and self efficacy. *Journal of Social & Clinical Psychology*, *25*(9), 875-884.
- Corrigan, P. W., & Calabrese, J. D. (2005). Strategies for assessing and diminishing self-stigma. In Corrigan, P. W. (Ed.). *On the stigma of mental illness: Practical strategies for research and social change* (pp. 239-256). Washington, DC, US: American Psychological Association.
- Cutrona, C. E., & Russell, D. W. (1990). Type of social support and specific stress: Toward a theory of optimal matching. In Sarason, B. R., Sarason, I. G. & Pierce, G. R.

- (Eds.). *Wiley series on personality processes. Social support: An interactional view* (pp. 319-366). Oxford, England: John Wiley & Sons.
- Faul, F., Erdfelder, E., Buchner, A., & Lang, A.-G. (2009). Statistical power analyses using G*Power 3.1: Tests for correlation and regression analyses. *Behavior Research Methods, 41*, 1149-1160.
- Field, A. (2005). *Discovering Statistics Using SPSS (Second Edition)*. London: Sage Publications.
- Field, A. (2013). *Discovering Statistics Using IBM SPSS Statistics (4th Edition)*. London: Sage Publications.
- Fritz, M., S., & MacKinnon, D., P. (2007). Required Sample Size to Detect the Mediated Effect. *Psychological Science, 18*(1), 233-239. doi: 10.1111/j.1467-9280.2007.01882.x.
- Ganzini, L., Johnston, W. S., & Hoffman, W. F. (1999). Correlates of suffering in amyotrophic lateral sclerosis. *Neurology, 52*(7), 1434-1434.
- Goffman E. (1963). *Stigma: Notes on a spoiled identity*. New Jersey: Prentice Hall.
- Goldstein, L. H., Atkins, L., Landau, S., Brown, R., & Leigh, P. N. (2006). Predictors of psychological distress in carers of people with amyotrophic lateral sclerosis: a longitudinal study. *Psychological Medicine, 36*(6), 865.
- Hayes, A. (2018). *Introduction to mediation, moderation and conditional process analysis: A regression-based approach (Second ed.)*. London: Guilford Press.
- Heijnders, M., & Van Der Meij, S. (2006). The fight against stigma: an overview of stigma-reduction strategies and interventions. *Psychology, health & medicine, 11*(3), 353-363.

- Henderson, C., Corker, E., Lewis-Holmes, E., Hamilton, S., Flach, C., Rose, D., ... & Thornicroft, G. (2012). England's time to change antistigma campaign: one-year outcomes of service user-rated experiences of discrimination. *Psychiatric Services, 63*(5), 451-457.
- Henry, J. D., & Crawford, J. R. (2005). The short-form version of the Depression Anxiety Stress Scales (DASS-21): Construct validity and normative data in a large non-clinical sample. *British Journal of Clinical Psychology, 44*(2), 227-239.
- Hogg, K. E., Goldstein, L. H., & Leigh, P. N. (1994). The psychological impact of motor neurone disease. *Psychological medicine, 24*(3), 625-632.
- Hunter, M. D., Robinson, I. C., & Neilson, S. (1993). The functional and psychological status of patients with amyotrophic lateral sclerosis: some implications for rehabilitation. *Disability and Rehabilitation, 15*(3), 119-126.
- Kenny, D. (2018). Moderator variables. Retrieved from <http://davidakenny.net/cm/moderation.htm#GO>
- King, R. C., Mulligan, P. K., & Stansfield, W. D. (2014). *A Dictionary of Genetics (8 ed.)* Oxford: Oxford University Press.
- Lazarus, R. S. (1966). *Psychological stress and the coping process*. New York: McGraw-Hill.
- Lazarus, R. S., & Folkman, S. (1984). *Stress, Appraisal, and Coping*. New York: Springer.
- Lillo, P., Mioshi, E., Zoing, M. C., Kiernan, M. C., & Hodges, J. R. (2011). How common are behavioural changes in amyotrophic lateral sclerosis?. *Amyotrophic Lateral Sclerosis, 12*(1), 45-51.

- Link, B. G., & Phelan, J. C. (2001). Conceptualizing stigma. *Annual Review of Sociology*, 27(1), 363-385.
- Levin, K. A. (2006). Study design III: Cross-sectional studies. *Evidence-Based Dentistry*, 7(1), 24.
- Lou, J. S., Reeves, A., Benice, T., & Sexton, G. (2003). Fatigue and depression are associated with poor quality of life in ALS. *Neurology*, 60(1), 122-123.
- Lovibond, P. F., & Lovibond, S. H. (1995). The structure of negative emotional states: Comparison of the Depression Anxiety Stress Scales (DASS) with the Beck Depression and Anxiety Inventories. *Behaviour research and therapy*, 33(3), 335-343.
- Lusli, M., Peters, R., van Brakel, W., Zweekhorst, M., Iancu, S., Bunders, J., & Regeer, B. (2016). The impact of a rights-based counselling intervention to reduce stigma in people affected by leprosy in Indonesia. *PLoS Neglected Tropical Diseases*, 10(12), e0005088.
- Maier, A., Holm, T., Wicks, P., Steinfurth, L., Linke, P., Münch, C., ... & Meyer, T. (2012). Online assessment of ALS functional rating scale compares well to in-clinic evaluation: a prospective trial. *Amyotrophic Lateral Sclerosis*, 13(2), 210-216.
- Matuz, T., Birbaumer, N., Hautzinger, M., & Kübler, A. (2010). Coping with amyotrophic lateral sclerosis: an integrative view. *Journal of Neurology, Neurosurgery & Psychiatry*, 81(8), 893-898.
- Matuz, T., Birbaumer, N., Hautzinger, M., & Kübler, A. (2015). Psychosocial adjustment to ALS: a longitudinal study. *Frontiers in psychology*, 6, 1197.

- McCluskey, L. F., Elman, L. B., Martinez-Lage, M., Van Deerlin, V., Yuan, W., Clay, D., ... & Trojanowski, J. Q. (2009). Amyotrophic lateral sclerosis–plus syndrome with TAR DNA-binding protein-43 pathology. *Archives of Neurology*, *66*(1), 121-124.
- McLeod, J. E., & Clarke, D. M. (2007). A review of psychosocial aspects of motor neurone disease. *Journal of the neurological sciences*, *258*(1-2), 4-10.
- Montes, J., Levy, G., Albert, S., Kaufmann, P., Buchsbaum, R., Gordon, P. H., & Mitsumoto, H. (2006). Development and evaluation of a self-administered version of the ALSFRS-R. *Neurology*, *67*(7), 1294-1296.
- Montgomery, G. K., & Erickson, L. M. (1987). Neuropsychological perspectives in amyotrophic lateral sclerosis. *Neurologic Clinics*, *5*(1), 61-81.
- Ngoc, T. N., Weiss, B., & Trung, L. T. (2016). Effects of the family schizophrenia psychoeducation program for individuals with recent onset schizophrenia in Viet Nam. *Asian Journal of Psychiatry*, *22*, 162-166.
- O'Brien, R. M. (2007). A caution regarding rules of thumb for variance inflation factors. *Quality & Quantity*, *41*(5), 673-690.
- Oh, J., & Kim, J. A. (2017). Supportive care needs of patients with amyotrophic lateral sclerosis/motor neuron disease and their caregivers: A scoping review. *Journal of clinical nursing*, *26*(23-24), 4129-4152.
- Orkibi, H., Bar, N., & Eliakim, I. (2014). The effect of drama-based group therapy on aspects of mental illness stigma. *The Arts in Psychotherapy*, *41*(5), 458-466.
- Orrell, R. W. (2016). Diagnosis and management of motor neurone disease. *The Practitioner*, *260*(1796), 17-21.

- Pagnini, F., Marconi, A., Tagliaferri, A., Manzoni, G. M., Gatto, R., Fabiani, V., ... & Palmieri, A. (2017). Meditation training for people with amyotrophic lateral sclerosis: a randomized clinical trial. *European Journal of Neurology*, *24*(4), 578-586.
- Pagnini, F., Phillips, D., Bosma, C. M., Reece, A., & Langer, E. (2015). Mindfulness, physical impairment and psychological well-being in people with amyotrophic lateral sclerosis. *Psychology & Health*, *30*(5), 503-517.
- Pfeiffer, P. N., Heisler, M., Piette, J. D., Rogers, M. A., & Valenstein, M. (2011). Efficacy of peer support interventions for depression: a meta-analysis. *General hospital psychiatry*, *33*(1), 29-36.
- Rao, D., Choi, S. W., Victorson, D., Bode, R., Peterman, A., Heinemann, A., & Cella, D. (2009). Measuring stigma across neurological conditions: the development of the stigma scale for chronic illness (SSCI). *Quality of life research*, *18*(5), 585-595.
- Rao, D., Elshafei, A., Nguyen, M., Hatzenbuehler, M. L., Frey, S., & Go, V. F. (2019). A systematic review of multi-level stigma interventions: state of the science and future directions. *BMC medicine*, *17*(1), 41.
- Rigby, S. A., Thornton, E. W., Tedman, S., Burchardt, F., Young, C. A., & Dougan, C. (1999). Quality of life assessment in MND: development of a Social Withdrawal Scale. *Journal of the Neurological Sciences*, *169*(1-2), 26-34.
- Scambler, G., & Hopkins, A. (1986). Being epileptic: coming to terms with stigma. *Sociology of Health & Illness*, *8*(1), 26-43.
- Simpson, J., McMillan, H., & Reeve, D. (2013). Reformulating psychological difficulties in people with Parkinson's disease: The potential of a social relational approach to disablism. *Parkinson's Disease*, 2013. <https://doi.org/10.1155/2013/608562>

- Simpson, J., & Thomas, C. (2014). Clinical psychology and disability studies: Bridging the disciplinary divide on mental health and disability. *Disability and Rehabilitation*, 37(14), 1299-1304.
- Strong M. J., Grace G. M., Orange J. B., Leeper H. A., Menon R. S. & Aere C. A. (1999). Prospective study of cognitive impairment in ALS. *Neurology*, 53, 1665 – 1670.
- Tedman, B. M., Young, C. A., & Williams, I. R. (1997). Assessment of depression in patients with motor neuron disease and other neurologically disabling illness. *Journal of the neurological sciences*, 152, s75-s79.
- Thoits, P. A. (1986). Social support as coping assistance. *Journal of consulting and clinical psychology*, 54(4), 416.
- Thomas, C. (2007). *Sociologies of disability, 'impairment', and chronic illness: Ideas in disability studies and medical sociology*. London: Palgrave.
- Thurman, T. R., Jarabi, B., & Rice, J. (2012). Caring for the caregiver: evaluation of support groups for guardians of orphans and vulnerable children in Kenya. *AIDS Care*, 24(7), 811-819.
- Uys, L., Chirwa, M., Kohi, T., Greeff, M., Naidoo, J., Makoae, L., ... & Holzemer, W. L. (2009). Evaluation of a health setting-based stigma intervention in five African countries. *AIDS Patient Care and STDs*, 23(12), 1059-1066.
- van der Beek, K. M., Bos, I., Middel, B., & Wynia, K. (2013). Experienced stigmatization reduced quality of life of patients with a neuromuscular disease: a cross-sectional study. *Clinical rehabilitation*, 27(11), 1029-1038.

- Vignola, A., Guzzo, A., Calvo, A., Moglia, C., Pessia, A., Cavallo, E., ... & Chiò, A. (2008). Anxiety undermines quality of life in ALS patients and caregivers. *European journal of neurology*, 15(11), 1231-1236.
- Worms, P. M. (2001). The epidemiology of motor neuron diseases: a review of recent studies. *Journal of the Neurological Sciences*, 191(1-2), 3-9.
- Zimet, G. D., Dahlem, N. W., Zimet, S. G., & Farley, G. K. (1988). The multidimensional scale of perceived social support. *Journal of personality assessment*, 52(1), 30-41.

Table 1: *Demographic information of participants (N= 77)*

Characteristic	<i>N</i>	%
Male	43	55.8
Female	34	44.2
Nationality		
Australian	8	5.8
British/English/Welsh/Northern Irish	18	23.4
Canadian	14	18.2
Dutch	1	0.7
German	1	0.7
Indian	1	0.7
Irish	3	2.2
New Zealander	18	23.4
NZ European	3	2.2
South African	4	2.9
Swedish	1	0.7
USA	2	1.4
USA Canadian	1	0.7
Unknown	2	1.4
Relationship status		
Single	3	2.2
Cohabiting/married/civil partnership	58	71.4
Divorced	9	6.5
Widowed	4	2.9

Other	3	2.2
Carer assistance required to complete survey		
Yes	7	9.1
No	70	90.9

Note. Participants were on average 59.1 years old ($SD = 10.6$; range = 36 - 83). Participants had an average time since symptom onset of 4.8 years ($SD = 4.4$; median = 3.5; range = 0.6 – 22). Participants had an average time since diagnosis of 3.5 years ($SD = 3.9$; range = 0.1 – 21).

Table 2: *Descriptive statistics for all study measures*

Variable	Mean	Standard Deviation	Cronbach's alpha
DASS-21 Stress	7.03	4.59	.86
DASS-21 Anxiety	5.32	3.85	.75
DASS-21 Depression	7.19	5.22	.91
SA-ALSFRS-R	29.00	8.62	.82
MSPSS Total	66.47	15.66	.94
MSPSS Significant Other	24.10	5.68	.94
MSPSS Family	22.16	6.16	.90
MSPSS Friends	20.21	6.27	.91
SSCI Total	32.83	16.12	.92
SSCI Self-Subscale	22.48	10.36	.90
SSCI Enacted-Subscale	10.09	7.09	.87

Note: **DASS-21** = Depression, Anxiety and Stress Scale (measuring psychological distress); **SA-ALSFRS-R** = Self-Administered Amyotrophic Lateral Sclerosis Functional Rating Scale Revised (measuring physical functioning); **MSPSS** = Multidimensional Scale of Perceived Social Support (measuring social support); **SSCI** = Stigma Scale for Chronic Illness (measuring stigma); SSCI Self-Subscale measures 'felt stigma' and SSCI Enacted-Subscale measures 'enacted stigma'

Table 3: *Correlation matrix of Spearman's correlation coefficients*

Variable	1	2	3	4	5	6	7	8	9	10
1. Age	1.000	.090	-.031	.198	-.134	-.140	-.179	-.175	-.026	-.052
2. Gender		1.000	-.165	-.097	.028	.014	-.016	.114	-.067	-.070
3. SA-ALSFRS-R			1.000	.255*	-.402**	-.395**	-.232*	-.153	-.209	-.180
4. MSPSS Total				1.000	-.483**	-.483**	-.433**	-.385**	-.399**	-.437**
5. SSCI Total					1.000	.929**	.801**	.538**	.447**	.660**
6. SSCI Self						1.000	.586**	.525**	.526**	.689**
7. SSCI Enacted							1.000	.440**	.244*	.465**
8. DASS-21 Stress								1.000	.627**	.787**
9. DASS-21 Anxiety									1.000	.588**
10. DASS-21 Depression										1.000

* $p < 0.05$; ** $p < 0.01$

Table 4: Mediation model of the indirect effect of MND related stress (enacted stigma) through felt stigma on psychological distress ($N = 77$).

Analyses	Point			Completely
	estimate	SE	95% CI	standardized effect
Depression Model				
Enacted Stigma → Felt Stigma (IV to mediator, path a)	.941***	.129	[.683, 1.198]	
Felt Stigma → Depression (mediator to DV, path b)	.316***	.053	[.211, .420]	
Enacted Stigma → Depression (total effect, path c)	.401***	.071	[.259, .543]	
Enacted Stigma → Depression (direct effect, path c')	.104	.077	[-.049, .257]	
Enacted Stigma → Depression (indirect effect, path $a \times b$)	.297↓	.061	[.191 - .429]	.404
Anxiety Model				
Enacted Stigma → Felt Stigma (IV to mediator, path a)	.941***	.129	[.683, 1.198]	
Felt Stigma → Anxiety (mediator to DV, path b)	.206***	.047	[.113, .300]	
Enacted Stigma → Anxiety (total effect, path c)	.196**	.058	[.080, .312]	

Enacted Stigma → Anxiety (direct effect, path <i>c'</i>)	.002	.068	[-.134, .138]	
Enacted Stigma → Anxiety (indirect effect, path <i>a x b</i>)	.194↓	.056	[.092 - .313]	.358

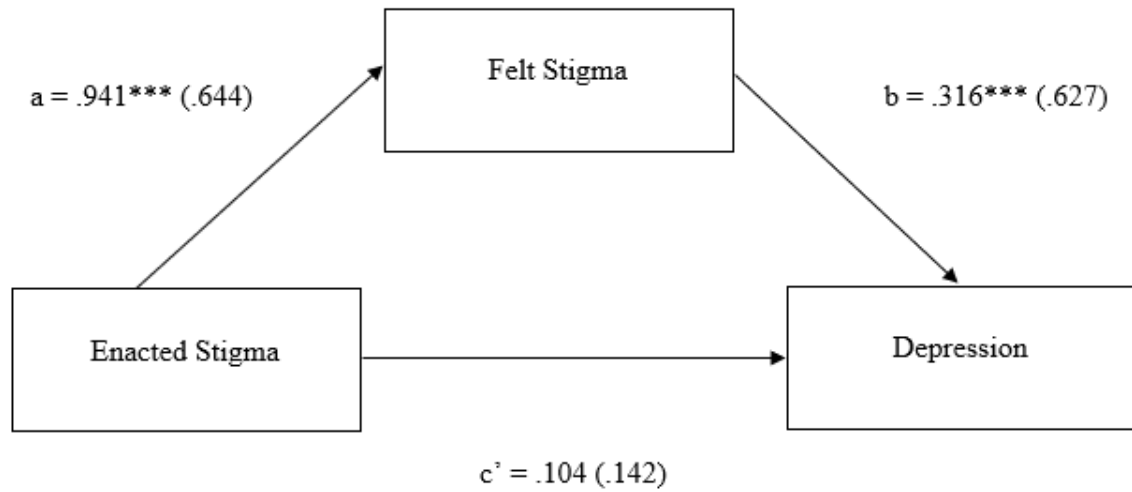
Stress Model

Enacted Stigma → Felt Stigma (IV to mediator, path <i>a</i>)	.941***	.129	[.683, 1.198]	
Felt Stigma → Stress (mediator to DV, path <i>b</i>)	.193***	.051	[.092, .294]	
Enacted Stigma → Stress (total effect, path <i>c</i>)	.365***	.062	[.242, .488]	
Enacted Stigma → Stress (direct effect, path <i>c'</i>)	.184*	.074	[.036, .332]	
Enacted Stigma → Stress (indirect effect, path <i>a x b</i>)	.182↓	.052	[.069 - .278]	.281

* $p < 0.05$; ** $p < 0.01$; *** $p < 0.001$

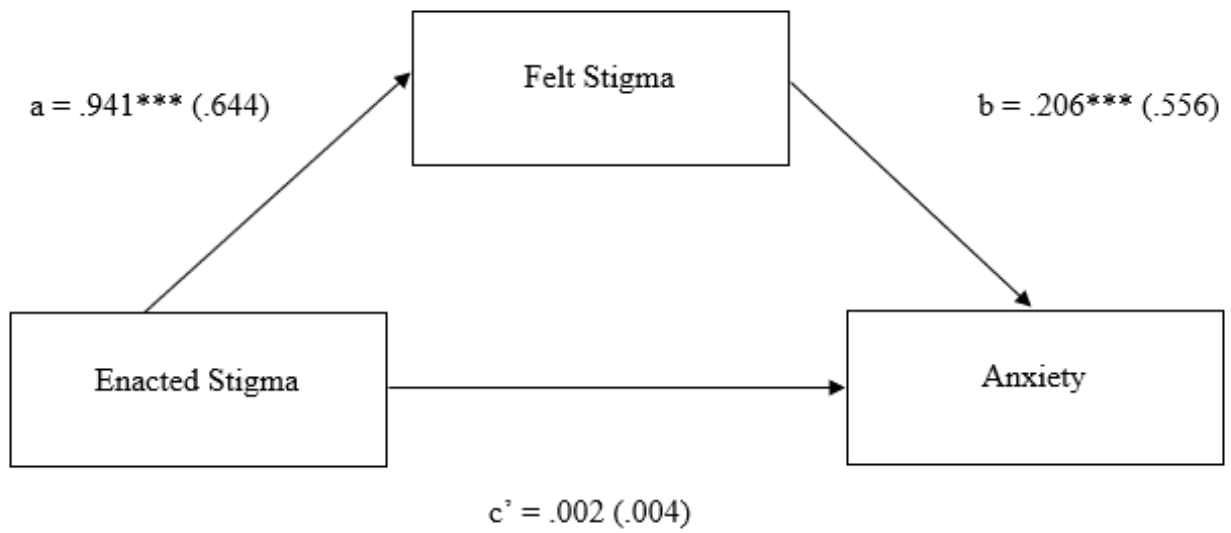
Note: ↓ - due to the way that the indirect effect is calculated, only a confidence interval, but not a p - value, is available.

Figure 1: Regression coefficients for the relationship between enacted stigma and depression as mediated by felt stigma. Standardized coefficients are included in parentheses.



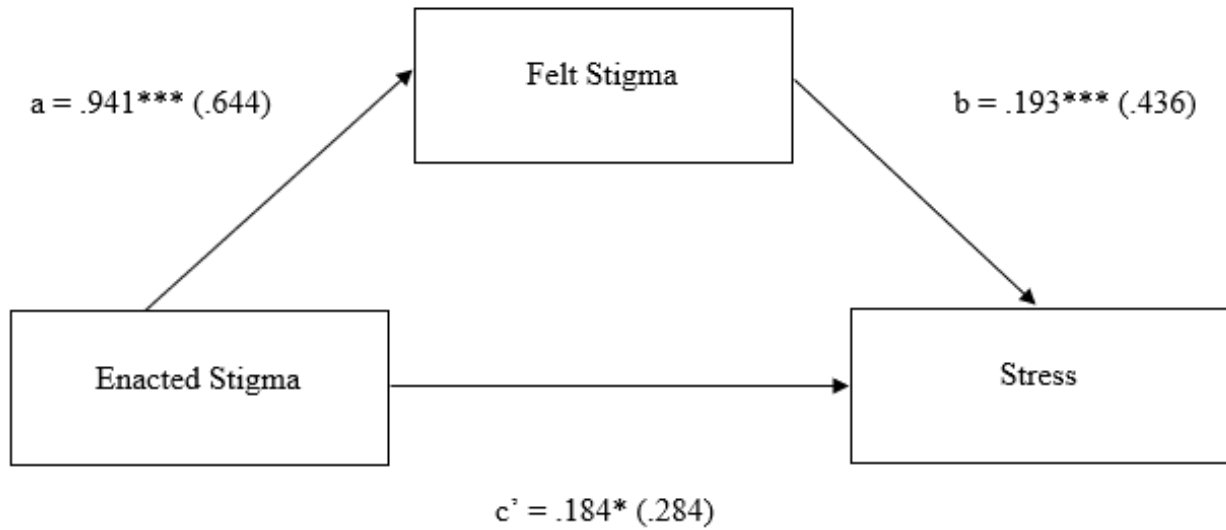
*** $p < 0.001$

Figure 2: Regression coefficients for the relationship between enacted stigma and anxiety as mediated by felt stigma. Standardized coefficients are included in parentheses..



*** $p < 0.001$

Figure 3: Regression coefficients for the relationship between enacted stigma and stress as mediated by felt stigma. Standardized coefficients are included in parentheses.



*** $p < 0.001$

* $p < 0.05$