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# Reducing Pneumonia Related Death in Amyotrophic Lateral Sclerosis Patients Through Improved Oral Health Care

Susan Wray Jones

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Susan W. Jones

*Candidate*

Dental Hygiene

*Department*

This thesis is approved, and it is acceptable in quality and form for publication:

*Approved by the Thesis Committee:*

*Christine Nafhe*

,Chairperson

*[Signature]*

*Demetra Logothetis*

*Eraine Morris*

**REDUCING PNEUMONIA RELATED DEATH IN  
AMYOTROPHIC LATERAL SCLEROSIS PATIENTS  
THROUGH IMPROVED ORAL HEALTH CARE**

**by**

**SUSAN WRAY JONES**

**B.S., Dental Hygiene, University of Nebraska, 1991**

**THESIS**

**Submitted in Partial Fulfillment of the  
Requirements for the Degree of**

**Master of Science  
Dental Hygiene**

**The University of New Mexico  
Albuquerque, New Mexico**

**May 2011**

## DEDICATION

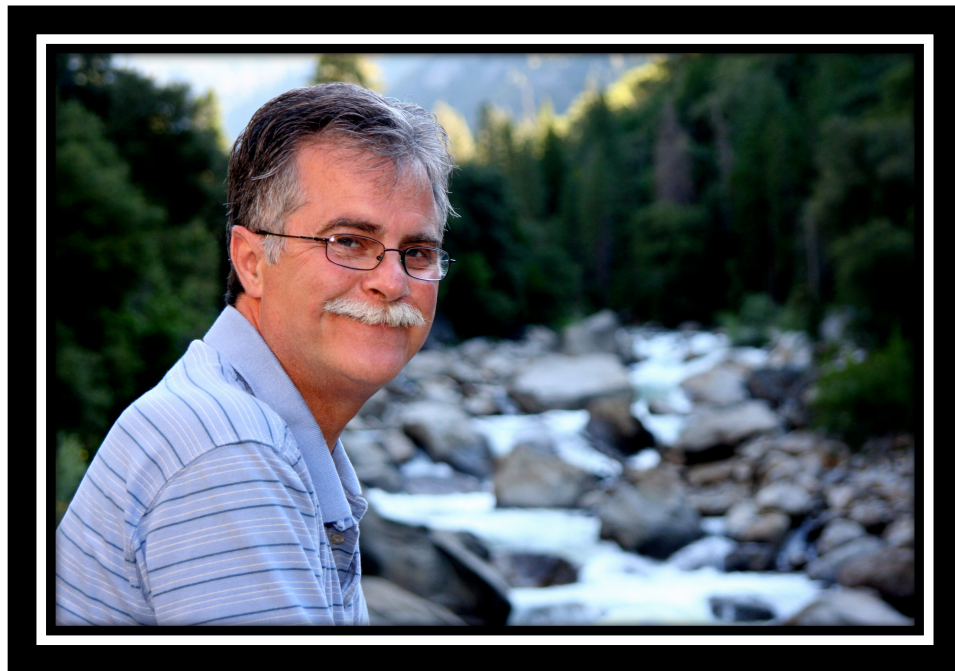
This thesis is dedicated to:

Those who have fought the ALS battle  
and  
Those who fought and must go on – their families

My three boys  
Darrín, Clayton and Colin  
Thank you for being my light ...there's never a wish better than this

and

In memory of Michael Allen Correll



“I have fought the good fight, I have finished the race, I have kept my faith...”  
2 Timothy 4:7

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To the collaborating ALS Association Certified Centers and ALS Association Executive Directors, thank you for your assistance with my research. I hope that the light shed upon this topic may, in some small way, positively impact the lives of those fighting the courageous and arduous battle with the monster that is amyotrophic lateral sclerosis.

To my parents, brother and sister, thank you for *always* being there.

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**ABSTRACT**

In patients diagnosed with Amyotrophic Lateral Sclerosis (ALS), the primary pathological cause of death, as determined by autopsy, is pneumonia. Research associates poor oral health with an increased incidence of pneumonia. The incidence is further elevated in mechanically ventilated, disabled and high-risk individuals. A proactive approach in oral health care could mitigate the risk of pneumonia related deaths in patients with ALS. This study evaluated whether the dental needs of ALS patients were being met.

Over a three-month period 121 ALS patients were surveyed regarding their oral health status. Both written and online survey formats were employed. ALS Association Multidisciplinary Clinics and Certified Centers assisted in survey promotion and distribution. Research results were analyzed utilizing bivariate Pearson correlation coefficients to determine relationships among study variables.

Results showed patients' difficulty in obtaining dental health care increased by 38.7% when the responsibility of their oral health care transitioned from patient to caregiver. Also increasing the difficulty in obtaining dental health care was the length of time elapsed since patient diagnosis. An overwhelming 85% of patients responded that they had not received dental health care information at their multidisciplinary clinic appointments.

Educating patients, their caregivers and ALS medical support personnel on the significance of oral health care and its association with pneumonia and pneumonia related death could have a positive impact on ALS patient life expectancy. This study supports the need for inclusion of a dental component into ALS patients' multidisciplinary clinic appointments.



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## Chapter 1: Introduction

Amyotrophic Lateral Sclerosis (ALS), also known as Lou Gehrig's disease, is a degenerative, fatal neuromuscular disease. Each year in the United States 5600 people are newly diagnosed. There is no known cure. From date of diagnosis typical life expectancy is three to five years. During that time the patient steadily deteriorates from a functional state into a completely paralyzed, fully dependent lifestyle. Pneumonia induced respiratory failure is the leading cause of death.<sup>1</sup> It is speculated that, as their disease advances, patients with ALS often find maintaining oral health quickly becomes a challenge. Recent studies associate poor oral hygiene and periodontal disease with colonized oropharyngeal respiratory pathogens that, when aspirated, may induce pneumonia.<sup>3</sup> Therefore, ALS patients with insufficient oral health and to an even higher degree, patients opting for mechanical ventilation are at an increased risk for developing life-threatening pneumonia. Research also supports the hypothesis that, in these susceptible patients, oral and dental health education can reduce the incidence of pneumonia.<sup>21</sup> As the disease progresses and oral health care responsibilities transition to the caregiver(s) dental hygiene education and instruction become pivotal to the patient's well-being.

### Statement of the Problem

As ALS patients' motor skills decline the level of difficulty in performing activities of daily living (ADL) and obtaining medical care increases. Not only is it an obstacle for the patient but, finances, transportation and treatment scheduling also

complicate the matter for caregivers. Dental health care and oral hygiene, initially maintained by the patient now become the responsibility of the caregiver.

Poor oral hygiene and periodontal disease have been positively linked with pneumonia, the leading cause of death in ALS patients.<sup>3</sup> Studies have also shown a reduction in pneumonia associated with poor oral health when preventative oral hygiene procedures are performed.<sup>3</sup> Therefore, it is imperative that the ALS patient, as well as their caregivers, understand not only the significance of oral health but are also trained in proper oral hygiene protocols. This research study was designed to determine whether the dental health care needs of ALS patients are being met.

### Significance of the Problem

There is no current research supporting the necessity of dental/oral health treatment during the progression of ALS. Nor is there research that identifies the varying levels of oral health care required by ALS patients throughout the course of the disease.

As the ALS patients' motor skills deteriorate, their ability to maintain oral health care also declines. Additionally, caregivers may not be aware of the significance of oral health in relationship to the specific needs of the patient. Oral hygiene instruction tailored to the various stages of the disease is easily disseminated; and the ALS patient's primary caregiver is in the optimal position to meet those needs.

Progressive weakening of the muscles utilized for eating, breathing, drinking and swallowing creates additional obstacles for the ALS patient.<sup>52</sup> Management of

these symptoms may include ventilatory support. Non-invasive bi-level positive airway pressure devices (or BiPAP®) machines are often incorporated at night for respiratory relief. Tracheostomy and direct mechanical ventilation devices are two additional treatment options, though seldom chosen by ALS patients.<sup>68,70</sup>

Regardless of the decision, all these options have profound effects on the oral cavity and one's ability to maintain dental health. Because the patient's condition may rapidly change, oral hygiene education needs to be provided to both the patient and the caregiver.

Obtaining dental care at a dental facility often poses its own set of complications. Not all dental practices are equipped to handle ALS patients in all of their variant stages. Patients may have to seek care from a different provider or forego dental treatment altogether. While changing health care providers can be stressful to any person, to the patient with ALS, this process may prove to be overwhelming and negatively influence their decision to seek care.

Evaluation of ALS patients' dental needs is imperative in providing optimal patient care. Assessing the patient's current dental condition and oral health knowledge may provide valuable information regarding whether the inclusion of a dental component within ALS Association Multidisciplinary Clinics would be beneficial.

### Operational Definitions

#### **Activities of Daily Living (ADL)**

Basic personal tasks in caring for oneself daily, for example, dressing, bathing and eating.

### **Amyotrophic Lateral Sclerosis (ALS)**

A chronic, progressive disease marked by gradual degeneration of the nerve cells in the central nervous system that control voluntary muscle movement. The disorder causes muscle weakness and atrophy. Symptoms commonly appear in middle to late adulthood, with death in three to five years. The etiology is unknown, and there is no known cure.

### **Upper Motor Neuron**

Found in the motor cortex area of the brain, they send messages to lower motor neurons to control skeletal muscle movement. Primarily involved with maintaining muscle tone and initiation of voluntary muscle movement.

### **Lower Motor Neuron**

Also known as anterior horn cells, they take messages from upper motor neurons to control the muscle fibers they innervate.

### **Bi-level Positive Airway Pressure (BiPAP)**

Mechanical ventilation designed to assist patients in moving air both into and out of lungs.

### **Mechanical Ventilation**

Machines used to assist or replace spontaneous breathing.

**Invasive** – Artificially supported ventilation provided through a cannula or breathing tube placed directly into patient's trachea.

**Non-invasive** – Ventilatory assistance delivered via a nasal, oral/nasal or full-face mask designed to replicate patients’ normal respiration.

### **Ventilator-Associated Pneumonia (VAP)**

Common complication causing significant morbidity and mortality in critically ill patients, broadly defined as pneumonia developing more than 48 hours after intubation.<sup>4</sup>

### **Durable Medical Equipment (DME)**

Adaptive, reusable medical equipment used to improve patient comfort and/or prolong patient independence. Must be medically necessary and prescribed by a physician.

### Assumptions

For the purpose of this investigation it is assumed that all patients surveyed have been previously, positively diagnosed with clinical evidence of amyotrophic lateral sclerosis and have provided honest, accurate responses regarding their condition.

It is also assumed that articles and documents referenced in this investigation utilized ethical and accurate methods to obtain and present factual information.

### Limitations

Despite numerous resources regarding amyotrophic lateral sclerosis, there was limited information available on the number of ALS patients that elect for mechanical ventilation, the type and extent of mechanical ventilation used and the

length of time between a patient's final clinic appointment and date of death.

Results indicating the number of ALS patients that choose to die at home in contrast to a skilled care facility and whether the role of primary caregiver was familial or an outside entity varied greatly, dependent upon the geographical region studied.

Initial survey results suggested time elapsed since patient diagnosis may help indicate the individual's current physical state. Collection of this information was included in the amended electronic survey. Clarification of the patient's degree of mobility and level of self-care at the time of survey may also prove valuable in future studies.

Although five ALS Certified Centers agreed to participate with written survey distribution and collection, the data from one location, Banner Good Samaritan Medical Center of Phoenix Arizona, was not received in time to be included in statistical analysis.

### Methodology

A research study was conducted in which 121 ALS patients were surveyed during a three-month period from November 2010 through January 2011. Patients were surveyed either in paper (written) format during routine ALS Association Certified Center appointments or a secure website. Previously diagnosed ALS patients who were 18 year of age or older were recruited. SPSS was employed for statistical analysis. The University of New Mexico Human Research Review Committee (HRRC), approved this study on Aug 16, 2010. Participant completion of survey implied consent.



## Chapter 2: Review of Literature

Literature review was conducted by researcher using medical subject headings (MeSH terminology) in PubMed/MEDLINE database from September 2010 – January 2011. Cited references from previously published articles were also used to locate additional resources.

### **MeSH terms:**

Pulmonary infection/disease

Respiratory

Amyotrophic Lateral Sclerosis

Periodontal disease

Oral health/hygiene

Intubation

Mechanical ventilation

Pneumonia

Oral/dental hygiene education

Oral/dental hygiene instruction

### **Key words:**

Respiratory, Amyotrophic Lateral Sclerosis, Periodontal, Oral hygiene, Pneumonia

### **Inclusion criteria:**

Articles that evaluated correlations between ALS, respiratory conditions, pneumonia, ventilation, dental/oral health and oral hygiene education/instruction were reviewed.

**Exclusion criteria:**

Search was limited to human studies.

**The following measures were assessed:**

Amyotrophic Lateral Sclerosis

Primary cause of death in ALS patients

Correlation between oral health and respiratory disease

Correlation between mechanically ventilated patients and respiratory disease

Reduction of respiratory complications through preventative oral health care, education and instruction

**Amyotrophic Lateral Sclerosis (ALS)****History**

It was Jean Martin Charcot, a French physician and founder of modern neurology who, in the 1870's, first linked the symptoms of amyotrophic lateral sclerosis to the group of nerves specifically affected by the disease.<sup>55</sup> It is believed Charcot employed a housemaid with ALS and was therefore able to systematically observe the clinical manifestations of her disease. His meticulous observations and descriptions of the disease remain accurate today. Throughout the world the disease is known by different names. In parts of Europe the disease is still known as Maladie de Charcot (Charcot's disease). In the United States the disease is widely recognized as ALS or Lou Gehrig's disease; named after New York Yankees baseball legend Lou Gehrig, who retired after being diagnosed in 1939. Until his death in 1941 Gehrig used his celebrity to promote awareness and educate the public about

the disease.<sup>64</sup> In 1993 the SOD1 gene on chromosome 21 was found to play a role in some cases of familial ALS.<sup>65</sup>

### **Epidemiology**

The annual incidence of ALS is one to two cases per 100,000 people. It is estimated that, during any given time, 30,000 people in the United States have the disease.<sup>9</sup> Each year an average of 5600 Americans are newly diagnosed, the equivalent of 15 people per day. The typical age of onset ranges from 40-70 years of age with the median age being 55. It is 20% more common in males and not affected by racial, ethnic or socioeconomic status.<sup>9</sup> From the date of diagnosis more than 50% will live three to five years. Approximately 20% will live more than five years, 10% more than ten years and 5% will live longer than twenty years.<sup>77</sup>

### **Etiology**

A multitude of theories have postulated the etiology of ALS. However, to date, none have proved statistically significant. Charcot first hypothesized a correlation between poliomyelitis and the development of ALS. In 1907, SAK Wilson questioned the roles of heavy metal involvement.<sup>11</sup> Wilson's theory gained momentum through the early 1900's until research showed mercury and lead, the leading suspects, were capable of producing reversible ALS-like symptoms. Since then, a host of prevailing theories including dietary deficiency, environmental factors, physical injury<sup>64</sup>, toxic exposure, vascular, immune and inflammatory disorders have been investigated without substantiation.<sup>52</sup> Current research leans toward a vast multifactorial approach encompassing genetics, viral, autoimmune and neurotoxic hypotheses.<sup>61</sup>

## **Clinical Features**

ALS is a disorder of the motor neurons responsible for contracting the skeletal muscles.<sup>11</sup> In health, lower motor neurons, located in the brain stem and spinal cord, innervate the muscle fibers. When a motor neuron fiber becomes diseased it can no longer control the muscle it innervates and denervation occurs. Consequently, the impaired muscle fibers shrink and the muscle becomes unable to contract. If surrounding motor neurons are healthy they can regenerate nerve fibers and take over control from the denervated fibers. Despite the overall reduction in the number of motor neurons, as long as this process is maintained, muscle strength remains constant.<sup>11</sup>

When the rate of denervation exceeds the rate of reinnervation, muscle strength decreases. In more than half of ALS patients the progressive weakening of muscles are among the first symptoms noted. Initial limb (arm or leg) weakness is more common than bulbar onset; which involves muscles controlling speech and swallowing.<sup>52</sup> Typical signs of lower motor neuron degeneration include muscle weakness, muscle atrophy, muscle cramping, fasciculations, paretic bulbar palsy (dysarthria, dysphagia, sialorrhea), hyporeflexia and difficulty swallowing which can lead to life-threatening aspiration issues.<sup>66</sup>

Upper motor neurons are located in the brain and brain stem. They carry information from brain centers that control the muscles of the body. These neurons synapse with the lower motor neurons which transmit information to the muscles.<sup>12</sup> Therefore, in order for motor commands generated in the central nervous system to reach the muscles of the body, signals must utilize both the upper and the lower

motor neurons. Upper motor neuron dysfunction progresses more slowly and reveals itself in symptoms such as loss of dexterity, muscle spasticity, hyperreflexia and spastic bulbar palsy.<sup>52</sup>

The type and degree of motor neuron onset are two of the main characteristics utilized in diagnosing and classifying ALS.<sup>52</sup> Another popular basis used in classification is the method of acquisition.<sup>52</sup> Ninety to ninety-five percent of patients have the sporadic form of ALS (SALS); ALS with no known familial history. The remaining 5-10% have familial ALS (FALS), with a close relative having the disease.<sup>65</sup> The clinical features of the two acquisition types are indistinguishable. In 1998 the El Escorial criteria was developed as the standard for classifying ALS patients in clinical research. These criteria are based on clinical evidence of lower and upper motor neuron involvement and represent the *degree* of diagnostic certainty.<sup>52</sup>

### **Diagnosis**

Currently there is no one hundred percent accurate, definitive test to diagnose ALS. Diagnosis is based on a myriad of signs, symptoms, neurological and laboratory data while simultaneously eliminating the diverse diagnostic alternatives that mimic ALS.<sup>52</sup> There multiple gene mutations associated with familial ALS, several of which can be tested by commercial labs.<sup>82</sup> The SOD1 genetic mutation represents the only known cause in ALS development.<sup>65</sup> However, subsequent studies on American and British FALS patients, have shown that the SOD1 genetic mutation only accounts for 20% of inherited ALS<sup>83,84</sup>

## **Treatment**

Predominant treatment consists of a variety of physical therapies and palliative drugs. In 1995, Rilutek® (riluzole) became the first FDA approved ALS drug.<sup>10</sup> To date, it remains the only FDA approved ALS drug available in the United States. Researchers believe riluzole helps protect the motor neurons from an overproduction of glutamate; a substance involved in nervous system function which, in excess, proves toxic to the neurons.<sup>76</sup> Clinical trials have demonstrated a modest two to three month extension in life expectancy.<sup>10,75,76</sup> In America ALS is considered an “orphan disease” in that it is a rare disorder affecting fewer than 200,000 people.<sup>10</sup> Consequently, drug discovery remains a challenge, as pharmaceutical companies are unlikely to make a profit from such a small market. Hence they do not dedicate resources to costly research and drug development. In 1983, Congress passed the Orphan Drug Act providing exclusive marketing rights and financial incentives to encourage orphan disease research.<sup>10</sup> Despite these initiatives, today more than 130 years later, ALS remains an incurable disease.

## **Primary Cause of Death**

Clinical death is defined as the cessation of the two primary criteria necessary to sustain life: blood circulation and breathing.<sup>5</sup> Most resources available on amyotrophic lateral sclerosis identify the leading cause of clinical death as respiratory failure.<sup>1, 6-8</sup> The pathological cause of death is diagnosed through laboratory analysis of bodily fluids and/or tissues.<sup>5</sup> In the post mortem state this process is known as an autopsy. To date, very little literature exists identifying the

cause of death in ALS patients through post mortem pathological analysis. It is estimated the autopsy rate among ALS patients is approximately four percent.<sup>6</sup> Within the body of research that does exist, the primary cause of death in more than 70 percent of the cases is either broncho-pneumonia or aspiration pneumonia.<sup>1,6,7,8</sup> A twenty-two year study following 100 ALS patients found a major discrepancy between the clinically assessed and pathologically determined cause of death.<sup>7</sup> A less than 20% concordance between clinical and pathological conclusions, showed clinical assessment not to be a reliable marker of death.<sup>7</sup>

### **Correlation Between Oral Health and Respiratory Disease**

Recent literature supports the association between respiratory disease and oral health and confirms the oral cavity to be a harbor of infectious respiratory pathogens.<sup>3</sup> In 2001, following a nine year study of 358 veterans, dental decay and the presence of cariogenic bacteria and periodontal pathogens were shown to be significant aspiration pneumonia risk factors.<sup>13</sup> Another study observed 189 elderly persons over a four year period and confirmed an association between pneumonia and decayed teeth. In this study dependence on caregivers was also linked to pneumonia.<sup>14</sup> A third study linked higher plaque scores with a previous history of respiratory tract infections.<sup>15</sup> Periodontal disease has been moderately associated with atherosclerosis, myocardial infarction and cardiovascular disease<sup>16</sup> and the risk of chronic obstructive pulmonary disease is also known to be significantly elevated with severe periodontal attachment loss.<sup>17</sup>

Pneumonia is defined as an inflammatory condition of the lung caused by bacterial, viral, fungal or parasitic infections.<sup>5</sup> The risk of pneumonia is, in part, determined by the specific bacteria inhaled and the body's ability to eliminate the bacteria from the airway mucosa. To eliminate aspirated bacteria from the lower airway, multiple defense mechanisms must function properly. Poor oral hygiene and the presence of periodontal disease may foster oropharyngeal colonization of respiratory pathogens which increase the probability of aspiration pneumonia, especially in high-risk patients.<sup>3,80</sup> The effectiveness of these mechanisms may also be further impaired by a variety of life conditions such as advanced age, residing in a nursing home or hospital and debilitated persons.<sup>3</sup>

### **Mechanical Ventilation and Respiratory Diseases**

In advanced stages of ALS the process breathing, or voluntarily exchanging oxygen and carbon dioxide, may become difficult and often results in respiratory distress. Symptoms often include headaches upon waking, physical exhaustion, labored breathing, and the inability to cough or speak long sentences. Decisions associated with ventilatory assistance become integral in ALS disease management.<sup>24</sup> Options include non-invasive positive pressure ventilation (NIPPV) and invasive ventilation. Both modalities demonstrate prolonged survival rates and are believed to improve the patient's overall quality of life.<sup>26-30</sup> Bilevel positive pressure airway machines, otherwise known as a BiPAP, are considered a non-invasive form of artificial ventilation. Unlike CPAP, which provides continuous airway pressure, the BiPAP unit provides two levels of air pressure, one on



inhalation and another on expiration. Variant nasal and facial delivery systems are available to assist with oxygen intake and carbon dioxide removal. There are other forms of non-invasive assisted ventilation available. However, in ALS patients, the BiPAP system is most prevalent.<sup>18</sup> Noninvasive positive pressure ventilation use in ALS patients has been associated with prolonged survival rates.<sup>18</sup> In recent years nocturnal NIPPV use has become the treatment of choice for ALS patients suffering chronic respiratory insufficiency.<sup>19</sup> The extent of NIPPV use in amyotrophic lateral sclerosis patients has not been widely studied.<sup>18</sup> One source estimated successful NIPPV use among ALS patients as high as 70% and suggested higher usage was likely limited by improperly fitted masks.<sup>68</sup>

Approximately three to five percent of ALS patients elected to have invasive ventilation.<sup>18,68</sup> The immediately aforementioned source also noted more frequent selection of invasive ventilation associated with cultural origin. For example, ALS patients residing in Japan more frequently select invasive ventilation than those residing in the United States.<sup>68</sup> Mechanical ventilatory support is provided via a tracheostomy, a surgically created opening in the trachea through which air is forced on an individualized, timed cycle.<sup>20</sup> This is an irreversible decision but may exponentially prolong the patient's life. Despite the benefit the cost of invasive ventilation is estimated between \$153K- \$336K per year. Additionally, invasive ventilation requires 24-hour skilled caregiver support.<sup>19</sup>

Nosocomial bacterial pneumonia, or ventilator-associated pneumonia (VAP), is the most common infection reported by intensive care units (ICU).<sup>2</sup> VAP is associated with a seven to twenty-one fold increase in the incidence of pneumonia<sup>2</sup>

and accounts for 47% of all ICU patient infections.<sup>32</sup> Twenty eight percent of mechanically ventilated patients will develop this pneumonia resulting in an attributable increase in morbidity and mortality.<sup>2</sup> Another study found nosocomial pneumonia prevalence in ICU's ranged from 10% – 65%, with a thirteen to fifty-five percent fatality rate.<sup>23</sup> Studies conducted in the 1990's suggest ventilator-associated pneumonia is a vital outcome determinant in critically ill patients.<sup>23</sup>

### **Reduction of Respiratory Complications Through Oral Health Care Measures**

Poor oral health can lead to complex medical and dental issues.<sup>31</sup> It has also been positively linked to an increased incidence of pneumonia in ventilated, debilitated and other high-risk patient groups.<sup>3,13</sup> A significant amount of research has been conducted, evincing consistent results, on the reduction of respiratory complications through oral hygiene measures. Standardized oral hygiene protocols can effectively decrease the colonization of dental bacteria associated with respiratory pathogens that cause pneumonia.<sup>33-39</sup> Mechanical toothbrushing and chlorhexidine mouthrinses were shown to be the two most effective modalities.<sup>33-39</sup> A 2009 study found, after implementation of an oral care protocol, a 46% reduction in ventilator-associated pneumonia cases during a twelve month period.<sup>36</sup> Studies confirmed these results not only with ICU patients but also in nursing home residents.<sup>34</sup>

The majority of ALS patients that elect to die home is variant upon geographical region. In Europe it approximately 52-63% of patients die at home in contrast to 85% electing to die at home within the United States.<sup>40-42,68,85</sup> One

source estimated 85% of ALS patients die under the care of a familial caregiver.<sup>68</sup> As the disease progressed, the caregiver's burden becomes enormous; exponentially more so when mechanical ventilation is involved. Many caregivers resort to hospice or hire skilled health care workers for assistance with the constant care that is required. Studies evaluating nurses' compliance of simple and CDC (Centers for Disease Control and Prevention) oral health care guidelines found that, in the ICU, these protocols were neither consistently nor uniformly implemented.<sup>32, 43-49</sup> Nurses were found to lack knowledge in recommended oral health care procedures<sup>44,45</sup> and discrepancies were noted among reported and actual practices and policies.<sup>48</sup> Despite significant evidence on the relationship between oral health care and disease prevention, nurses generally fail to fully appreciate its implications.<sup>49</sup>

### Chapter 3: Methods and Materials

This study's conclusions were based on the results from 60 paper surveys distributed to existing ALS patients at ALS Association Certified Center appointments and 61 ALS patients participating in the survey online. Participant completion of the survey implied consent. The survey was anonymous and no personal identifiers were collected.

ALSA Certified Center Medical Directors were sent initial contact letters inviting their participation. Five of the thirty-four clinics contacted participated. Certified Center Medical Directors were asked to sign and return participation commitment letters. Contact information regarding their Center's designated

survey coordinator was also collected. Participants then received a survey packet containing 27 patient surveys with attached information cover letters.

Designated individuals were asked to distribute surveys to ALS patients, or their primary caregiver, over the age of eighteen. As all patients attending Certified Center multidisciplinary appointments had been previously diagnosed with amyotrophic lateral sclerosis; therefore, patient privacy and survey recipient selection was not an issue. Patient cover letters described the content of the survey, investigator contact information, Institutional Review Board (IRB) approval, informed consent and the purpose of the study. Location markers were pre-printed on each survey and used only to identify only the site from which the data were collected. On a pre-determined end date, ALS Association Certified Center designated individuals returned all completed surveys.

Flyers promoting the identical survey in an online format were mailed to ALS Association Executive Directors in November 2010. Directors were asked to distribute flyers to ALS patients over age 18. ALS Association Executive Directors disseminated flyers to ALS patients throughout their respective regions of the United States promoting the online survey. In an attempt to prevent duplicate responses, flyers were not sent to regions participating in the written survey. Directors were free to disseminate flyers in electronic or paper format. SurveyMonkey, a web-based interface specializing in the creation and publishing of custom web surveys, was utilized for the online effort. Parameters were set to prohibit multiple responses from a unique IP address. The online data collection period ran from November 2010 through January 2011.

Demographic data including gender, onset diagnosis, type of ALS, age at diagnosis, history of military service, intubation and ventilatory support were collected. To determine whether patients had an increase in dental treatment needs, patient root canal therapy and amalgam dental restorations were assessed both prior to and since diagnosis. Participants were also questioned on the frequency and difficulty in obtaining dental care prior to and since diagnosis. The primary person responsible for performing oral hygiene care as well as the routinely implemented dental hygiene regimens was also asked. Patients were questioned as to whether they had received oral hygiene education or instruction at their ALS Association Multidisciplinary Center appointments. A final section allowed patients to add qualitative comments they felt were pertinent to the study.

#### Chapter 4: Results

The study sample was compromised of 121 diagnosed amyotrophic lateral sclerosis patients over the age of eighteen.

Population demographics:

Gender

Gender	Male	Female	Did not identify
N =	61	56	4

Survey population age range

Age range	18-29 years	30-39 years	40-49 years	50+ years	Not disclosed
N =	2	5	25	87	2

History of military service

Yes 21%; No 79%

### ALS Onset Type

75 participants reported bulbar onset, 80 participants limb onset.

16 respondents did not identify onset type.

### Form of ALS

7% (n=8) reported familial form ALS (FALS)

73% (n=88) reported sporadic form ALS (SALS)

21% (n=25) did not identify ALS form.

### Current Oral Health Care Regimes

94% toothbrushing

44% flossing

12% swabbing

53% mouthrinse

7% utilize other mechanisms

### Data Analysis

A statistical analysis of the data was formatted in the SPSS 11.5 statistical software program and utilized standard bivariate correlations, using Pearson correlation coefficient, to determine the level of inter-dependence among variables. Relevant data was analyzed using standard frequency counts, cross tabulations (measuring inter-variable relationships) and stepwise regression.

At the 95% statistical confidence level, the bivariate correlation analysis showed a strong correlation between variables measuring increased difficulty in obtaining professional dental care and 1) presence of a feeding tube, 2) patient's

inability to eat food through mouth and 3) caregiver(s) as primarily responsible for dental health care.

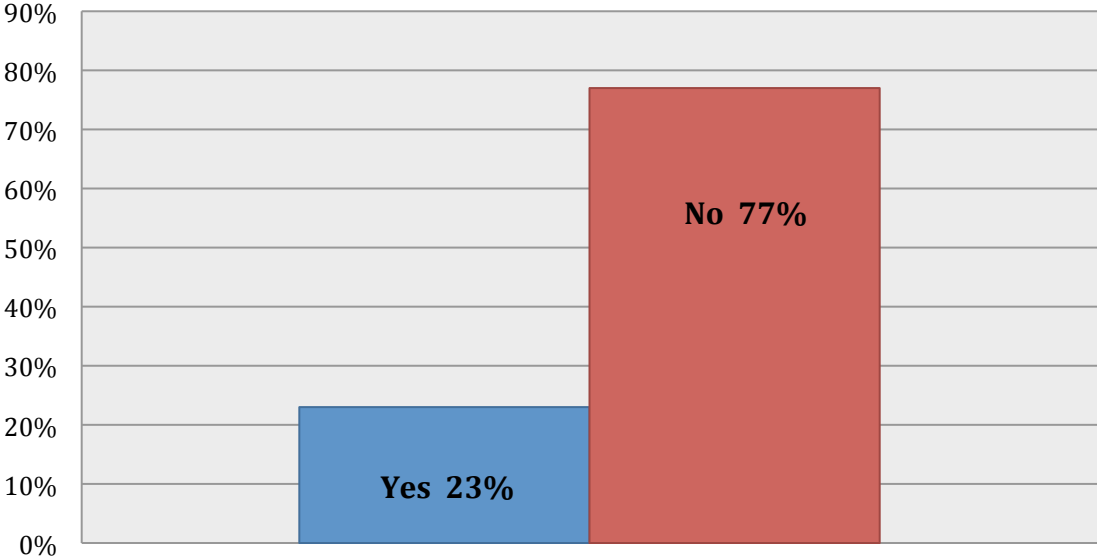
### Cross Tabulations

		Intubation	Feeding Tube	Oral Food Intake	Noted Decreased Frequency of Dental Visits	Increased Difficulty in Obtaining Dental Care	Person Responsible For Dental Care
Intubation	Pearson Correlation	1	.494**	-.287**	.035	.156	-.336**
	Sig. (2-tailed)		.000	.002	.712	.100	.000
	N	114	112	110	111	112	113
Feeding Tube	Pearson Correlation	.494**	1	-.607**	→.175	→.301**	-.468**
	Sig. (2-tailed)	.000		.000	.064	.001	.000
	N	112	117	113	113	114	115
Oral Food Intake	Pearson Correlation	-.287**	-.607**	1	-.171	-.195*	.288**
	Sig. (2-tailed)	.002	.000		.072	.038	.002
	N	110	113	116	112	113	114
Noted Decreased Frequency of Dental Visits	Pearson Correlation	.035	.175	-.171	1	.188*	-.168
	Sig. (2-tailed)	.712	.064	.072		.043	.070
	N	111	113	112	117	117	117
Increased Difficulty in Obtaining Dental Care	Pearson Correlation	.156	.301**	-.195*	.188*	1	-.299**
	Sig. (2-tailed)	.100	.001	.038	.043		.001
	N	112	114	113	117	118	118
Person Responsible For Dental Care	Pearson Correlation	-.336**	-.468**	.288**	-.168	→-.299**	1
	Sig. (2-tailed)	.000	.000	.002	.070	.001	
	N	113	115	114	117	118	119

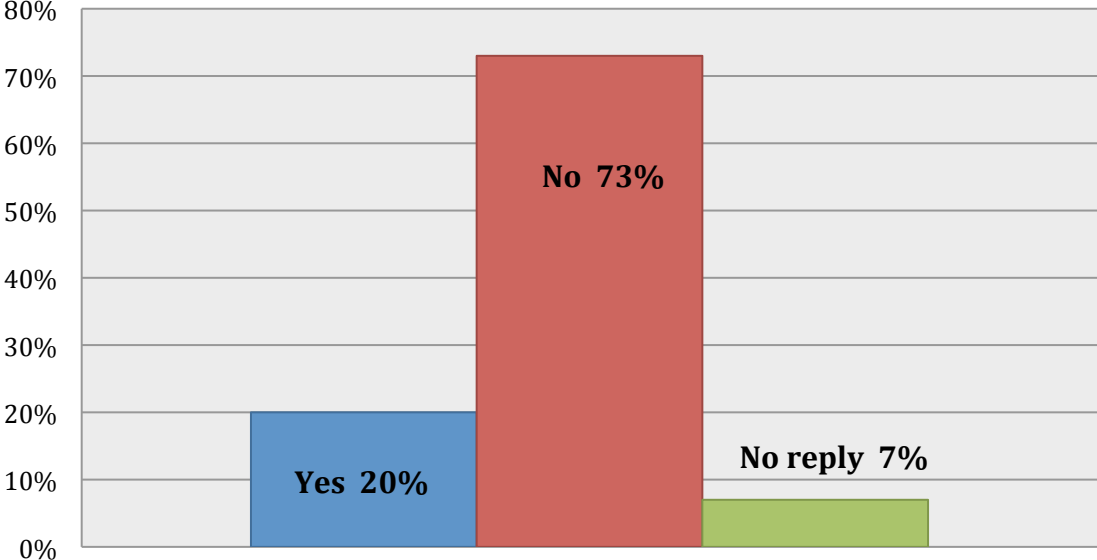
\*\* . Correlation is significant at the 0.01 level (2-tailed).  
 \* . Correlation is significant at the 0.05 level (2-tailed).

Frequency analysis of the same variables showed 23% of respondents have feeding tubes, 26% are under the primary care of a caregiver, and 20% of these individuals find it increasingly difficult to obtain professional dental care. Strong existing correlations between these variables suggest that the majority of these characteristics are associated with the same ALS patients.

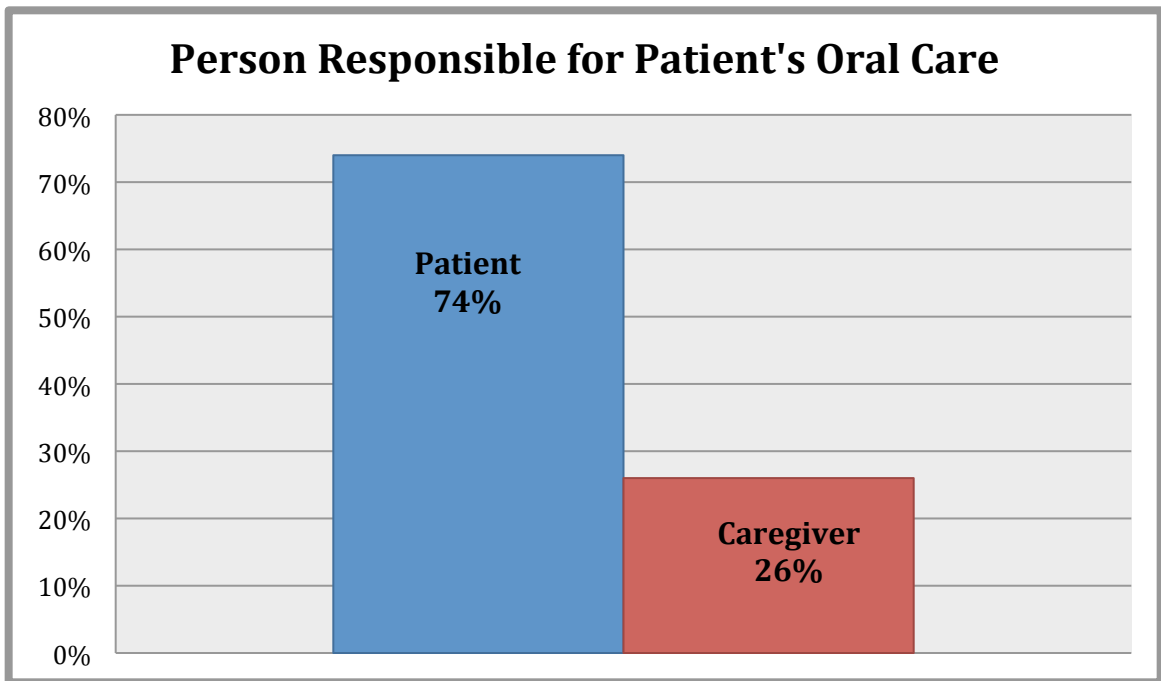
### Currently Have Feeding Tube



### Increased Difficulty in Obtaining Dental Care







Of those ALS patients under the responsibility of a primary caregiver, 38.7% reported an increase in difficulty obtaining dental care, compared to 13.8% of patients capable of self-care. This finding suggests that as the disease progresses and the responsibility of dental health care transitions to a caregiver, the difficulty in obtaining dental health care increases. Oral health instruction and education may be warranted amongst both groups.

**Increased Difficulty in Obtaining Dental Care \* Person Responsible for Dental Care  
Cross Tabulation**

			Person Responsible For Dental Care		Total
			Self	Caregiver	
Increased Difficulty in Obtaining Dental Care	Yes	Count	12	12	24
		% within Person Responsible For Dental Care	13.8%	38.7%	20.3%
	No	Count	67	19	86
		% within Person Responsible For Dental Care	77.0%	61.3%	72.9%
	Don't Know	Count	8	0	8
		% within Person Responsible For Dental Care	9.2%	.0%	6.8%
Total		Count	87	31	118
		% within Person Responsible For Dental Care	100.0%	100.0%	100.0%

The increase in difficulty in obtaining dental care variable also showed a direct relationship with the length of time since diagnosis response. As the disease ascends to the three-year mark, the likelihood of the patient experiencing increased difficulty accessing dental care also increases; a logical conclusion given the average time elapsed from diagnosis to death is three to five years.

**Increased Difficulty in Obtaining Dental Care \* Length of Time Since Diagnosis  
Cross Tabulation**

			Time Since Diagnosis Groups					Total
			Less Than 1 Year	1-3 Years	3-5 Years	6-10 Years	10+ Years	
Increased Difficulty in Obtaining Dental Care	Yes	Count	1	6	5	3	1	16
		% within Time Since Diagnosis Groups	8.3%	22.2%	45.5%	42.9%	25.0%	26.2%
	No	Count	9	19	5	4	3	40
		% within Time Since Diagnosis Groups	75.0%	70.4%	45.5%	57.1%	75.0%	65.6%
	Don't Know	Count	2	2	1	0	0	5
		% within Time Since Diagnosis Groups	16.7%	7.4%	9.1%	.0%	.0%	8.2%
Total		Count	12	27	11	7	4	61
		% within Time Since Diagnosis Groups	100.0%	100.0%	100.0%	100.0%	100.0%	100.0%

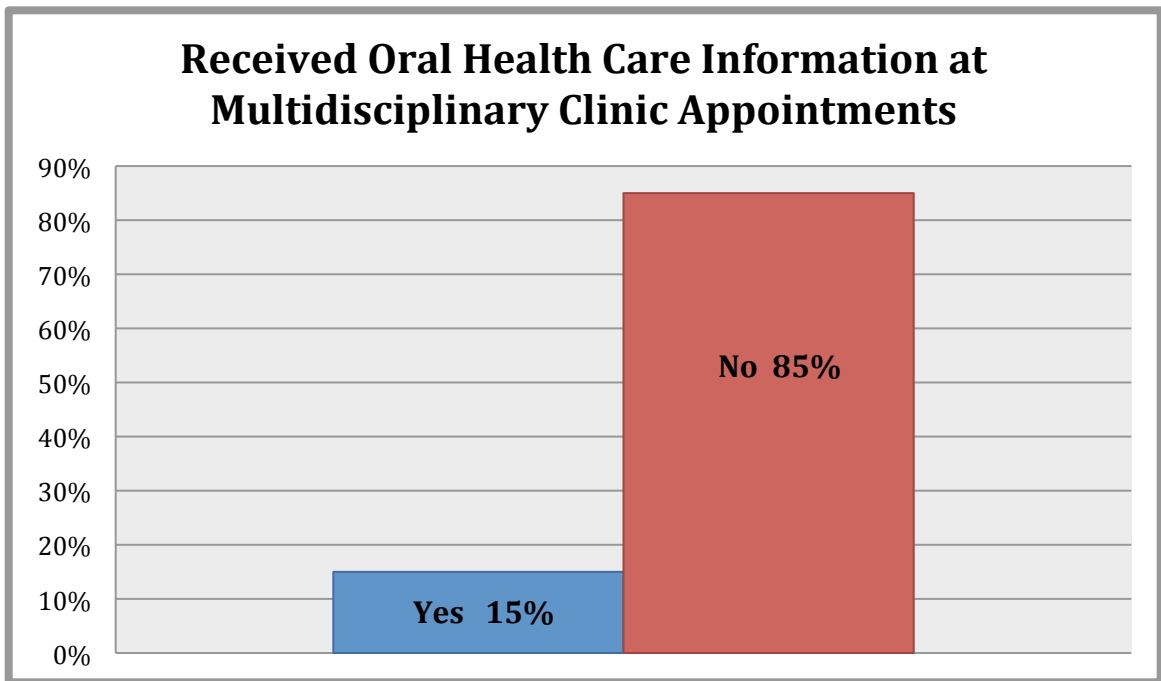
Further analysis of patients with feeding tubes, also indicating increased difficulty in obtaining dental care, showed a trend in the number of silver/metal, (amalgam) fillings present prior to diagnosis in relation to the likelihood of having a

feeding tube. 10.5% of respondents with 1-3 fillings prior to diagnosis had a feeding tube; that number increased to 21.1% in those with 4-6 fillings and 52.6% among those with 7 or more fillings. While these results indicate a potential trend, the sample size of those with feeding tubes and prior fillings, 19 respondents, was statistically small.

**Number of Fillings \* Presence of Feeding Tube - Cross Tabulation**

			Feeding Tube		Total
			Yes	No	
Number of Fillings	1 to 3	Count	2	18	20
		% within Feeding Tube	10.5%	25.4%	22.2%
	4 to 6	Count	4	24	28
		% within Feeding Tube	21.1%	33.8%	31.1%
	7+	Count	10	16	26
		% within Feeding Tube	52.6%	22.5%	28.9%
	Don't Know	Count	3	13	16
		% within Feeding Tube	15.8%	18.3%	17.8%
Total	Count		19	71	90
	% within Feeding Tube		100.0%	100.0%	100.0%

Frequency analyses revealed an overwhelming 85% of respondents have not received information regarding dental health care at their ALS Association Multidisciplinary Clinic Appointments. This finding strongly advocates the inclusion of a dental component in amyotrophic lateral sclerosis patient's multidisciplinary appointments.



CHAPTER 5 – Discussion

This study surveyed 121 patients over the age of eighteen diagnosed with ALS. The original intent was to survey patients at all of the 34 ALS Association Certified Centers. However, many of the ALS Association Certified Center Medical Directors were reluctant to participate without obtaining Institutional Review Board (IRB) approval from their respective institution. Of the 34 centers invited to participate, five gave consent. During a telephone conversation with Dr. Richard S. Bedlack, Medical Director of Duke ALS Clinic, on 9/29/2010<sup>80</sup> he suggested distribution of a flyer promoting the survey in an online format might produce better compliance. An online survey was created; utilizing SurveyMonkey.com, and amended HRRC (IRB) approval was obtained from the University of New Mexico. Then, letters were sent to 57 regional ALS Association Chapter Executive Directors. The survey results obtained from this method of contact proved not only easier in

promotion and distribution but also in data collection. The demographics of this sample population strongly resembled overall ALS population characteristics regarding gender, age, type of ALS onset, history of military service and disease form.

In order to expand the scope of the study several additional dental related parameters were included. These questions inquired about the type and number of dental restorations present in the patient's mouth before and since diagnosis. The two survey formats differed only in that the online survey included an additional question, asking patients the length of time since their diagnosis. Seventy-four percent of the overall sample patient population responded that they were capable of self-care. In earlier stages of ALS when patients are less likely to be compromised with mobility and swallowing difficulties, it was anticipated patients' overall level of self-care would not only be higher but, that their oral health care regimen might also include normal activities of self care such as brushing and flossing. With disease progression it was anticipated oral health care responsibilities would transition to a caregiver and obtaining professional dental health care would become more difficult. Results from the online survey population, where length of time since diagnosis was reported, strongly supported these correlations. The results suggested that oral health care regimens among patients where a greater amount of time had elapsed since diagnosis were represented in the 12% that reported swabbing in their dental care. While swabbing alone does not provide effective oral health care, the addition of an antimicrobial agent such as chlorhexidine has shown significant improvement in patient periodontal pocket depths as well as plaque and calculus levels.<sup>78</sup>

Inclusive of the entire survey population, a strong correlation existed between the presence of a feeding tube, the inability to eat food through the mouth, and caregiver(s) having primary responsibility of the patient's oral health care. This suggested that as the disease advanced and nutritional intake through the mouth became more challenging, patients were more likely to have feeding tubes and their daily oral health care regimen dependent on a caregiver(s). Percutaneous Endoscopic Gastrostomy (PEG), or feeding tubes, are an effective system used to provide patients with nutrition and hydration.<sup>71</sup> Used in ALS patients, PEG's, have been shown to stabilize the weight loss typically experienced.<sup>71</sup> However, it is also important to recognize, even though the patient is not participating in traditional oral food intake, their need for dental health care does not decrease. When dental health care is neglected, bacterial pathogens within the oral cavity continue to thrive. If not disrupted through oral health care measures, the already at-risk patient possesses an even greater increased risk of developing life-threatening pneumonia.<sup>3</sup>

Almost 40% of respondents whose care is dependent on a caregiver reported an increased difficulty in obtaining professional dental health care. In the early stages of ALS, patients are more likely to be capable of self-care and maintaining routine professional dental care appointments. Within these appointments, patient education by the dental professionals regarding the significance of oral health care throughout the course of their disease is pivotal. As the overall health status of the patient decreases other complications in seeking professional dental care arise. Patient mobility, transportation, treatment scheduling, accessibility to dental offices with wheelchair accommodations as well as symptoms of dysphagia, dyspnea,

variant saliva levels, muscle spasticity, fatigue and other obstacles further impede access to traditional dental care. If patients are able to obtain professional dental care while under the supervision of a caregiver, caregiver oral health education and instruction at these appointments becomes even more critical. Research clearly indicates that even trained critical care nurses, with specific oral health guidelines, do not consistently comply with recommended oral hygiene protocols.<sup>43-46</sup> This supports the necessity of understanding the significance of oral health care as essential to the ALS patient's well-being, not only by the patient but also the caregiver, who may or may not be a trained skilled care worker.

Data from the online survey also suggested a relationship existed between the length of time since diagnosis and difficulty in obtaining dental care. The correlation between difficulty in obtaining dental care and ALS onset diagnosis type was .034; although this leaned slightly toward limb onset, it was not a statistically significant association. As ALS patients migrate away from individualized medical appointments and toward multidisciplinary clinic appointments, the significance of oral health care education and instruction is often minimized if recognized at all. Over 85% of the total survey population reported not having received dental health care information at their multidisciplinary clinic appointments. If the patients had not been educated at previous professional dental care appointments, it is likely they do not fully appreciate the association between poor oral health and the incidence of pneumonia as well as the measures integral in mitigating these risks.

Significant research has been conducted regarding mechanically ventilated and other high-risk patients; this research advocates the use of oral hygiene

protocols to reduce the overall incidence of pneumonia.<sup>33,36-39</sup> Oral decontamination with 2% chlorhexidine solution alone has been strongly linked to a reduction in bacterial colonization and incidence of ventilator associated pneumonia (VAP) among mechanically ventilated patients.<sup>35</sup> Limited research exists on the prevalence of ventilator use among ALS patients, though it has been estimated that BiPAP® use may be as high as 70%.<sup>68</sup> Non-invasive positive pressure ventilation patients (BiPAP®) are also considered at high-risk for acquiring pneumonia due to fluctuating salivary levels, swallowing, mobility difficulties and other oral health care considerations.<sup>80</sup>

Currently, very little information exists regarding the significance of oral health care in amyotrophic lateral sclerosis patients. While ALS Association multidisciplinary clinics are in an optimal position to provide these patients and their caregivers with critical oral health care information and education, 85% of survey respondents reported this aspect had been overlooked. The ALS Association website does include a section entitled “Oral Care for the Patient with ALS: A Guide for the Caregiver,” however, it does not address the association between poor oral health and increased risk of pneumonia. Nor does it address the use of antimicrobial agents in mitigating this risk.<sup>79</sup> Education on the significance of oral health care and its association with pneumonia and pneumonia related death could be easily distributed to patients and caregivers in informative pamphlets at clinic appointments or website links. In later disease stages, oral swabbing with antimicrobial agents such as chlorhexidine could be implemented at a minimal cost. The ALS patient and their caregivers do have numerous other oral health care



options ranging from modified manual and electric toothbrushes to highly effective suction devices as well as the antimicrobial agents already discussed. A dental hygienist could provide valuable information during education and dental screenings at ALS Association chapter meetings, local support groups and multidisciplinary care visits. In many cities mobile dental units are available to assist in both routine and critical dental health maintenance venues.

Some of the most interesting and informative data within the survey results were the qualitative patient comments. Upon reading, it would be difficult to argue that dental care has not been neglected. Patients and caregivers alike were confused regarding the type of dental treatment needed as well as how and where to obtain it. Respondents posed questions concerning the need for a general dentist versus a specialist and whether an association existed between silver/metal “amalgam” fillings and the acquisition of ALS. Individual responses also included “difficult to brush,” “choking,” and “biting lip and inside jaw”. To a health care professional, these responses were difficult to hear, especially knowing we can educate and assist these patients during their time of greatest need.

It has been estimated over 80% of amyotrophic lateral sclerosis patients in the United States die within their own home under the primary care of a familial caregiver.<sup>1,42,45,68</sup> Current research clearly shows, upon post mortem examination, the leading causes of death in ALS patients are aspiration and broncho-pneumonia.<sup>7</sup> Recognizing the correlation between poor oral health and pneumonia, and the even greater risk in those patients utilizing mechanical ventilation, one can appreciate the significance of oral health care information and education in prevention of

pneumonia related death.<sup>13,33-39</sup> Studies evaluating the knowledge and compliance level of high-risk and intensive care unit nurses uncovers an alarming discrepancy between recommended oral hygiene procedures and actual performance protocols.<sup>43-50</sup> Due to the substantial cost and 24-hour care required for ALS patients, families are often forced to rely on a variety of caregivers rather than a single or limited number of skilled care personnel. If these medical professionals are not cognizant of the correlation between poor oral health and risk of pneumonia, how can we expect the average ALS patient or their caregivers to understand the importance of an effective oral health care regimen? In conclusion, recognizing 85% of patients reported not having received oral or dental health care information at their ALS Association multidisciplinary clinic appointments, it is logical to assume the majority of patients with amyotrophic lateral sclerosis and their caregivers do not fully appreciate the significance of a proper oral health care regimen. This demonstrates the necessity for inclusion of a dental health care component into ALS patient multidisciplinary clinic appointments. Few documented studies have been conducted on the oral health status of ALS patients throughout the course of their disease. This study has validated the fact that there are many integral components within the relationship between ALS and overall oral health. Future research should include a prospective study evaluating the extent and specific types dental health care interventions needed.

## APPENDIX A

### Actual Qualitative Patient Survey Comments

*Please add any additional comments that pertain to your experience with ALS and Dental Considerations: (Open-Ended Response)*

1. During a 30 day stay in 2 different hospitals I found dental hygiene to be the most overlooked aspect of the care I received
2. Lost filling this week, should it be put back in?
3. It is hard to hold the toothbrush as hands are weak and tire quickly. Fingers getting weaker so much harder to floss, especially in back - and it hurts to open my mouth wide. Why did you ask about filling and root canals?
4. Started using a power toothbrush 6 mos ago.
5. choking episodes at dentist
6. I receive all my care including dental care through the VA.
7. when I went for my last cleaning and exam, I asked the hygienist to keep my head higher than usual and they were happy to comply. they have another patient with ALS that they are working with to accommodate his needs
8. Dr. Jeffrey Day of Fredericksburg, Va. accepted me w/o any hesitation. He is very mindful of my ALS

9. My wife is totally bedridden. She is unable to even sit in a wheelchair. A little over a year ago my wife experienced severe teeth grinding. She broke the bone holding her front bottom six teeth. They were loose and wobbly and we were afraid that she would aspirate her teeth. Our dentist came to the house and removed those teeth. Since then several other teeth have "disappeared." We used to brush her teeth and suction out the toothpaste, but we now swab her teeth with 50% peroxide and water daily.
10. I have PLS.
11. I lost 3/4 of my teeth at age 18, the remainder at age 65
12. My dentist is not especially sympathetic to my needs. I tell him that I do not want x-rays because I choke on the pieces that you put in your mouth and his response is something like "well, we need to take x-rays to spot problems with your teeth." I tell him that I have bigger problems than my teeth, but it's in one ear and out the other.
13. handicap accessibility, dentists willingness to deal with the difficulties of ALS dental care, dental offices ability to accommodate the power wheel chair in their spaces have all made dental care difficult for my mother. I have found that county dental clinics are the most receptive to her with her condition....private practices

have an array of reasons why they will not accept her...not always sure how real those reasons are....suspect they just don` t want to have to deal with the more complex client.

14. Biting my lip and inside of jaw.
15. none
16. Should we see a Special Dentist? Or Regular one?
17. Professional dental care not practical due to high risk of aspirating liquids
18. gagging is an issue
19. Have used "Water Pic" both before & after onset.
20. Since I thought I had a limited time left I admit I thought one thing I could do without was dental cleaning. Wrong! After not having any cavities as an adult I now have two small ones and I had a decalcified spot repaired. So it is back to the dentist I am going. My dental office was marvelous about making things as easy for me as possible. For instance I did not have to change rooms - cleaning, exams, repair, x-rays, and payment processing were all done in same room. And there is a back exit for handicap patients to come and go.
21. My wife is very concerned as to whether or not amalgam fillings containing mercury could be causing ALS in people. Is there enough evidence that the amalgam fillings should be removed?

22. Difficult to locate a dentist that can accommodate a power chair.
23. I made my first visit to a dental facility at a VA hospital last week. I have to wait 6 wks. to get an approval from the VA to get outside help with my teeth.
24. did not go to the dentist before diagnoses no need. still have not had a need to see dentist
25. i have noticed breathing difficulties when laying prone in a dental chair. My dentist has been very accommodating in making my dental visit as easy and comfortable as possible.
26. Harder to brush
27. No dental coverage
28. Had trouble circling (responses) wife is a dental hygienist
29. Had dentures 20+ years
30. My wife is a hygienist
31. ...patient receives cleanings from dentist every three months (prior to diagnosis, every six months) patient has difficulty opening mouth wide enough during professional cleanings, making effective dental care challenging

## APPENDIX B

### SurveyMonkey Patient Flyer ALS Association Executive Director Participation Invitation

The first time I met “Ben” I was working as a dental hygienist in a private practice dental office. Ben was newly diagnosed with Amyotrophic Lateral Sclerosis. We cleaned his teeth, talked to him about his home care and sent him home like every other patient. But as you know Ben was not like every other patient. It was not until my Uncle was diagnosed with ALS that I fully understood the complexity of ALS and the obstacles in obtaining dental health care.

Oral health care is an integral component to every person’s physical well being, including those diagnosed with Amyotrophic Lateral Sclerosis (ALS). It is speculated that as the disease advances patients with ALS often find maintaining and receiving oral health care can quickly become a challenge.

Elaine Sanchez Dils and Susan Jones with the University of New Mexico Department of Surgery, Division of Dental Hygiene are conducting a research study investigating dental considerations in ALS patients. We would like to include the patients that visit the ALS Centers in your region.

Patients, or their designated individual, would be handed a flyer asking them to participate in an anonymous online survey about their dental experiences since diagnosis. The survey takes about 5 minutes to complete and could be completed at the location and time of their choice. Research study information is listed below.

The flyer is attached for printing and distribution. If you prefer hard copies, please contact us and we will mail them directly to you. The end date for data collection is January 31, 2011. Please feel free to contact us with any questions.

Thank you for your time and assistance,

Elaine Sanchez Dils, RDH, MA  
Associate Professor  
Principal Investigator  
(505)272-0838  
[EDils@salud.unm.edu](mailto:EDils@salud.unm.edu)

Susan W. Jones, RDH, MSc  
Master’s Degree Candidate  
Research Coordinator and Co-Investigator  
(505)272-4513  
[SWJones@salud.unm.edu](mailto:SWJones@salud.unm.edu)

University of New Mexico IRB Approved 8/16/2010 (HRRC# 10-200)  
Federal Wide Assurance Number: 00003255

Survey link:

<https://www.surveymonkey.com/s/ALSdentalresearchstudy>

**APPENDIX C**  
**SurveyMonkey Patient Flyer**

*Seeking ALS patients to participate in research study*

**You can make a difference...**



**my Husband**  
May 1966 -

**my Uncle**  
Sept 1953 - May 2009

**my Grandfather**  
Aug 1916 - July 1982

**and You**

**...for us as well.**

The University of New Mexico is conducting a research study to determine whether or not the dental needs of ALS patients are being met.

***You can help.***

**To participate go to:**

<https://www.surveymonkey.com/s/ALSdentalresearchstudy>

**Survey is anonymous and takes about  
5 minutes to complete.**

Please participate by January 31, 2011

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Thank you for your time.

Please contact us with any questions.

Elaine Sanchez Dils RDH, MA, Associate Professor  
edils@salud.unm.edu  
505-272-0838

Susan W. Jones, RDH, Masters Degree Candidate  
swjones@salud.unm.edu  
505-272-4513

(IRB/ HRRC #10-200. Approved 16-August-2010)



APPENDIX D

SurveyMonkey Patient Survey  
(Questions numbered in survey logic format)

Dental Considerations in Patients with Amyotrophic Lateral Sclerosis (ALS)

1. Default Section

\* 1. Please list the ALS Certified Center where you received this survey flyer

2. Age at time of ALS diagnosis

3. Length of time since diagnosis

4. Gender

Male

Female

5. ALS onset diagnosis

Bulbar onset

Limb onset

6. Type of ALS

Familial

Sporadic

7. Do you have a history of military service

Yes

No

8. Are you intubated?

Yes

No

9. Do you have a feeding tube?

Yes

No

10. Can you eat food through your mouth?

Yes

No

**Dental Considerations in Patients with Amyotrophic Lateral Sclerosis (ALS)**

**11. Prior to diagnosis did you have any silver(amalgam) tooth fillings in your mouth?**

- Yes
- No
- Not known

## Dental Considerations in Patients with Amyotrophic Lateral Sclerosis (ALS)

2.

**1. Approximately how many silver (amalgam) fillings were in your mouth prior to ALS diagnosis?**

- 1-3
- 4-6
- 7 or greater
- Not known
- Not applicable

**2. Prior to diagnosis did you have root canal therapy on any teeth?**

- Yes
- No
- Not known

## Dental Considerations in Patients with Amyotrophic Lateral Sclerosis (ALS)

3.

**1. Approximately how many root canal therapies were done prior to ALS diagnosis?**

- 1-3
- 4-6
- 7 or greater
- Not known

**2. Prior to diagnosis how frequently did you receive professional dental care?**

- Only with pain
- Rarely
- 2 or more times per year
- Not known

**3. Since diagnosis how frequently have you received professional dental care?**

- Only with pain
- Rarely
- 2 or more times per year
- Not known

**4. Since diagnosis has the frequency of your dental care decreased?**

- Yes
- No
- Not known

**5. Who is responsible for performing your daily oral home care (brushing, flossing, etc.)?**

**Check all that apply**

- Myself
- Caregiver
- Other

## Dental Considerations in Patients with Amyotrophic Lateral Sclerosis (ALS)

**6. What daily home care regimes are performed regularly?**

**Check all that apply**

Brushing

Flossing

Swabbing

Rinses

Other

**7. Since diagnosis have you seen an increase in difficulty in accessing professional dental care?**

Yes

No

Not known

**8. Have you received information regarding oral health or dental care during your ALS Association Certified Center appointments?**

Yes

No

**9. Please add any additional comments that pertain to your experience with ALS and Dental Considerations:**

## APPENDIX E

### ALS Association Certified Center Medical Director Participation Invitation Letter

The ALS Clinic at Penn State Milton S. Hershey Medical Center Department of Neurology, EC037  
30 Hope Drive  
Hershey, PA 17033

Dear Dr. Simmons,

Dental health care is an integral component to every person's physical well being, including those diagnosed with Amyotrophic Lateral Sclerosis (ALS). Routine dental examinations are critical in ensuring that existing and subsequent dental concerns do not progress into complications. It is speculated that as their disease advances patients with ALS often find maintaining and receiving oral health care quickly becomes a challenge. An evaluation of patients with ALS dental considerations since their diagnosis would provide valuable insight into whether or not these patients could benefit from the inclusion of a dental component into their existing ALS Association Certified Center appointments.

Elaine Sanchez Dils and Susan Jones with the University of New Mexico Department of Surgery, Division of Dental Hygiene are conducting a research study investigating dental considerations in ALS patients. With your permission we would like to include your patients. On August 16<sup>th</sup>, 2010 the University of New Mexico Institutional Review Board granted approval to this study (HRRC 10-200). The Federal Wide Assurance Number for this survey is: FWA 00003255. Therefore, it is not required for your Institution to obtain IRB approval unless you so desire.

Patients, or their designated individual, would be asked to complete a survey about their dental experiences since diagnosis. The survey would be distributed to patients at their previously scheduled ALS Association Certified Center appointments. The survey takes approximately 5 minutes to complete and could therefore be completed during practitioner transition times. Surveys would then be placed into a sealed envelope and returned to an appointed clinic location or employee. On the designated end date (December 1, 2010), all surveys would be returned in a provided parcel to the investigators.

As follow-up reminders may be necessary, surveys contain an identification number for institutional tracking purposes only. To ensure confidentiality surveys are anonymous and results will be used in summary format only. Steps have been taken to simplify the survey distribution and collection procedure. One of which includes a cover letter for each survey. In this form it states that "... completion of the survey implies consent". Therefore, identifiers or signatures from faculty, staff and patients would not be required on any paperwork.

As part of the confirmation of your institution's involvement, it is asked that the attached Center Participation letter be signed and returned via postal or e-mail. The letter verifies your institution's sole responsibility in this project in distributing and collecting surveys. It also confirms that you and your institution would not have other study interaction. Upon receipt of your returned letter, we will send the package containing all necessary components.

These is not a funded research project and are therefore unable to extend monies to staff member(s) who help with survey distribution and collection. It is our hope that the ALS Certified Centers involved with the study will choose to participate for the future enrichment of their patient's well being.

We appreciate your consideration and your timely response. If you have any questions please contact either investigator.  
Sincerely,

Elaine Sanchez-Dils, RDH, MA  
Associate Professor  
[EDils@SALUD.UNM.EDU](mailto:EDils@SALUD.UNM.EDU)  
505-272-0838

Susan W. Jones, RDH, BS  
Masters Degree Candidate  
[SWJones@SALUD.UNM.EDU](mailto:SWJones@SALUD.UNM.EDU)  
505-272-4513

#### Attachments (3)

1 – Survey Cover Letter/Patient Consent; 2 – Patient Survey; 3 – Center Participation Consent Letter

## APPENDIX F

### ALS Association Certified Center – Medical Director Participation Consent

(Recipient Name)  
Director, ALS Association Certified Center  
(School Name)  
(Street Address)  
(City, ST Zip code)

University of New Mexico  
Division of Dental Hygiene  
2320 Tucker NE, MSC09 5020  
(505) 272-4513

Research Investigators,

This letter is to confirm that (Insert Clinic Name) is willing to allow our patients, diagnosed with Amyotrophic Lateral Sclerosis to participate in the “Dental Considerations in Patients with Amyotrophic Lateral Sclerosis” research study.

I am aware that by signing this letter I am acknowledging our institution and any of its employees will have the sole responsibility of distributing and collecting the anonymous surveys. It also confirms that our institution and its employees will not have further interaction with the research study.

We do ask if the primary point of contact will not be you, the Director of the ALS Association Certified Center, you provide below the name, telephone number, physical address and e-mail address for the individual you would like the investigators to directly contact.

I understand that if I have questions regarding the study or this letter, I may contact the Principal Investigator, Elaine Sanchez Dils (505-272-0838 – [EDils@salud.unm.edu](mailto:EDils@salud.unm.edu)), Research Coordinator and Co-Investigator, Susan Jones (505-272-4513 – [SWJones@salud.unm.edu](mailto:SWJones@salud.unm.edu)) or the University of New Mexico Health Sciences Center Human Research Review Committee (505-272-1129).

It is the understanding and expectation that the above named research study will only commence upon complete approval from the University of New Mexico Health Sciences Center Human Research Review Committee.

Sincerely,  
(Recipient Name)  
Director, ALS Association Certified Center  
(Phone number)  
(E-mail address)

#### **Alternate Contact Person Information**

Name:  
Telephone Number:  
Address:  
E-mail Address:

## APPENDIX G

### ALS Patient Survey – Informed Consent Cover Letter

University of New Mexico Health Sciences Center

Informed Consent Cover Letter for Anonymous Surveys

#### STUDY TITLE

#### Dental Considerations in Patients with Amyotrophic Lateral Sclerosis

Professor Elaine Sanchez Dils RDH, MA and Susan Jones RDH, BS from the Department of Surgery, Division of Dental Hygiene, are conducting a research study. The purpose of the study is to determine whether or not the dental needs of Amyotrophic Lateral Sclerosis (ALS) patients are adequately being met. You are being asked to participate in this study because of your ALS diagnosis.

Your participation will involve completion of a short written survey. The survey should take about 5 minutes to complete. Your involvement in the study is voluntary, and you may choose not to participate. There are no names or identifying information associated with this survey. The survey includes questions such as "Since diagnosis how frequently has patient received dental care?" You can refuse to answer any of the questions at any time. There are no known risks in this study, but some individuals may experience discomfort when answering questions. All data will be kept for 5 years in a locked file in Professor Sanchez Dils' office and then destroyed.

The findings from this project will provide information on whether or not ALS patients would benefit from the inclusion of a dental component to their ALS Association Certified Center appointments. If published, results will be presented in summary form only.

If you have any questions about this research project, please feel free to call Elaine Sanchez Dils at (505)272-0838 or Susan Jones at (505) 272-4513. If you have questions regarding your legal rights as a research subject, you may call the UNMHSC Office of Human Research Protections at (505) 272-1129.

By returning this survey in the envelope provided, you will be agreeing to participate in the above described research study.

Thank you for your consideration.

Sincerely,

Researcher's Name & Title

Elaine Sanchez Dils, RDH, MA  
Associate Professor

Susan Jones, RDH, BS  
Masters Degree Candidate

HRRC#10-200

Version Date: 4/9/10

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APPROVED	8/16/10	
The University of New Mexico Human Research Review Committee		



**APPENDIX H**  
**Dental Considerations in Patients with Amyotrophic Lateral Sclerosis (ALS)**  
**Patient Survey**

**Location Code:**

**Gender of Patient:** M      F  
**Age at time of ALS Diagnosis:** \_\_\_\_\_  
**ALS Onset Diagnosis:**      Bulbar Onset              Limb Onset  
**Type of ALS:**              Familial              Sporadic  
**Does you have a history of military service?**    Yes    No

**Please circle answer:**

1. Are you intubated?	Yes	No			
2. Do you have a feeding tube?	Yes	No			
3. Can you eat food through your mouth?	Yes	No			
4. <b>Prior</b> to diagnosis, did you have silver (amalgam) tooth fillings in your mouth?	Yes	No	Not known		
5. <b>Proceed to next question if you answered 'NO' or 'NOT KNOWN' to question #5.</b> Approximately how many silver (amalgam) fillings were in your mouth prior to diagnosis?	1-3	4-6	7 or greater	Not known	
6. <b>Prior</b> to diagnosis, did you have root canal therapy on any teeth in your mouth?	Yes	No	Not known		
7. <b>Proceed to next question if you answered 'NO' or 'NOT KNOWN' to question #6.</b> Approximately how many root canal therapies were done in your mouth prior to diagnosis?	1-3	4-6	7 or greater	Not known	
8. <b>Prior</b> to diagnosis, how frequently did you receive professional dental care?	Only with pain	Rarely	2 or more times per year	Not known	
9. <b>Since</b> diagnosis, how frequently have you received professional dental care?	Only with pain	Rarely	2 or more times per year	Not known	
10. <b>Since</b> diagnosis, has the frequency of professional dental care decreased?	Yes	No	Not known		
11. <b>Since</b> diagnosis, have you seen an increased difficulty accessing regular dental care?	Yes	No	Not known		
12. Who is responsible for performing your daily home oral care (brushing, flossing, etc)?	Patient	Caregiver	Other		
13. What daily home care regimes are performed regularly ( <b>circle all that apply</b> )?	Brushing	Flossing	Swabbing	Rinses	Other
14. Have you received information regarding oral health or dental care during your ALS Association Certified Center appointments?	Yes	No			

\*Please add any additional comments that pertain to patient experiences with ALS and Dental considerations.

## **APPENDIX I Study Collaborators**

Medical Director, Zachary Simmons, M.D.  
Research Coordinator, Helen (Beth) Stephens M.A, CCRC  
The ALS Clinic at Penn State Milton S. Hershey Medical Center  
Hershey, Pennsylvania

Medical Director, Sarah Youssof, MD  
ALS Center University of New Mexico  
Albuquerque, New Mexico

Medical Director, Kevin Felice, D.O.  
Assistant Research Director, Lavanya Rajachandran Ph. D  
Neuromuscular/ALS Clinic  
Hospital for Special Care  
New Britain, Connecticut

Medical Director, Ezgi Tiryaki, MD  
Research Coordinator, Cindy Rhode, RN  
Hennepin Faculty Associates, Multispecialty Clinic  
Minneapolis Medical Research Foundation  
Minneapolis, Minnesota

Medical Director, Todd Levine, M.D.  
Banner Good Samaritan Medical Center  
Phoenix, Arizona

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