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Physician perceptions about living organ donation in patients with Amyotrophic Lateral Sclerosis

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Abstract

Objectives—Patients with Amyotrophic Lateral Sclerosis (ALS) have expressed desire to become living organ donors but are unable to do so with current organ donation policies. Our objective is to assess ALS patient's interest in organ donation, and perceived concerns of this practice by ALS neurologists.

Patients and Methods—An electronic survey was administered to ALS neurologists across the United States regarding living organ donation in ALS patients prior to respiratory failure.

Results—52 complete responses were received from 121 invites. 67% (35/52) of neurologists expressed no concerns about living organ donation in ALS patients, and 33% had concerns. The concerns related to respiratory failure, anesthesia exposure and renal dysfunction. With their concerns addressed, 71% of neurologists reported that they would endorse living organ donation.

49% of neurologists reported being asked by a patient for information regarding living organ donation. ALS neurologists felt that 22.8% of ALS patients (median 19%) would be interested in learning more about organ donation, while only 6% of neurologists broach this subject with their patients.

Conclusion—Our results indicate that 1 in every 4 ALS patients may be interested in exploring options for living organ donation, and this topic is not routinely addressed by ALS clinics. These results indicate an unexplored area of patient interest. To honor a patient's wishes to donate, the transplant community will have to accommodate living organ donation from terminally ill patients, and address neurologist concerns. Such a practice could benefit two groups of patients.

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Search terms

Amyotrophic lateral sclerosis; living organ donation; kidney donation

Introduction

Organ transplantation is a widely practiced intervention in patients with terminal organ failure since the first successful organ transplant in humans performed in 1954[1]. In 2015 alone, 30,974 organs (17,898 kidneys) were transplanted in the United States[2]. During the same period, a staggering 119,778 patients remained on the United Network for Organ Sharing (UNOS) waiting list, including 99,238 patients waiting for kidneys[2,3]. The vast majority of organs (88%) are recovered from deceased donors, while a lesser percentage are from living organ donors. Given the need for transplantable organs and the higher quality of organs with living donation, there have been recent campaigns to increase living or "good Samaritan" organ donation, which is strongly supported by public opinion[4]. The current UNOS policy for living organ donation dictates that potential donors be adults in good overall physical and mental health, and thus excludes all potential donors with chronic illness[5].

Recent media reports have highlighted multiple patients diagnosed with a terminal disease who have exhibited a strong desire to participate in living organ donation[6–9]. Of note, all of these patients carry the diagnosis of Amyotrophic Lateral Sclerosis (ALS). ALS is a fatal neurodegenerative condition in which the overwhelming majority of clinical disease is motor[10]. A small subset of patients have, to varying degrees, a frontotemporal lobe pattern of cognitive dysfunction, but the majority retain decision making capacity[10]. These patients face an incurable disease and expected mortality due to respiratory failure within 3 years of symptom onset, prolonged only with tracheostomy, ventilation and aggressive life support[10,11]. Many such patients find solace in the idea of organ donation to cure others with terminal disease, prior to respiratory failure and end of life[6–9]. However, living organ donation by ALS patients is not supported by transplant centers and federal policies, and to the author's knowledge has not been carried out.

We designed and administered a survey to ALS neurologists to assess the level of interest amongst them and their patients regarding living organ donation, prior to respiratory failure and end of life. We also enquired about potential concerns from the ALS neurologist community towards this idea.

Materials and Methods

An electronic survey was designed which consisted mostly of binary (yes/no) and multiple-choice style questions with some options for free response. The survey questionnaire is available in the supplementary appendix of this manuscript (supplement 1). Research Electronic Data Capture (Redcap™) hosted at the University of Utah, was used to implement the survey[12]. REDCap™ is a secure, web-based application designed to support research, data collection and manipulation.

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The authors compiled a list of ALS neurologists in the United States and a survey invite was sent electronically. Survey respondents were informed that “living organ donation is defined as donation of a single kidney, while the donor is alive, and that organ recovery surgery involves general anesthesia and typically lasts 1-2 hours.” Survey responses were managed, and statistical calculations done using REDCap™ and Microsoft Excel™.

Results

132 survey invites were sent and with 11 returned emails (due to incorrect contact information), the potential the total respondent list was 121. A total of 52 responses were received and included in the final analysis, with a response rate of 42.9%. Respondents were asked to provide a postal code of the location of their ALS practice and this geographic data is presented in Figure 1.

Graphical flowchart representation of the survey results is depicted in Figure 2. 5.9% (3 of 51) of ALS neurologists reported routinely discussing organ donation with their patients (deceased donation). 67.3% (35 of 52) reported no concerns with living organ donation in this patient population. The 32.7% (17 of 52) with concerns were asked to choose from different categories of concerns (more than one option could be chosen), as well as the ability to write in concerns. 9 of the 17 neurologists had concerns about the ALS patient/donor, 12 of the 17 had concerns related to the organ recipient, and 1 wrote a free response that stated that local state laws would exclude ALS patients from organ donation (the survey respondent was located in the state of Georgia).

Of the 9 neurologists (out of 17 with concerns) who reported safety concerns for the ALS donor patient, 7 opted to specify their concerns as: risks of prolonged ventilator dependence following anesthesia (7 of 7), exposure to anesthetic agents (3 of 7), and risk of uropathy or renal dysfunction with a single kidney (2 of 7).

Within the12 neurologists (out of 17) that reported concerns regarding the organ recipient, all focused on the possible transmission of disease to the organ recipient. Possible transmission vectors were: ALS-associated protein (12 of 12), ALS-associated virus (8 of 12), ALS-associated gene (4 of 12), and the age of the recipient for possible transmission (3 of 12). The 17 respondents with concerns were asked if they would support living organ donation if their concerns were addressed, and 76.5% (13 of 17) expressed support.

49% of ALS neurologists (25 of 51) reported being spontaneously asked by at least one patient, for information and options related to living organ donation. When neurologists were asked to estimate the percentage of ALS patients who would be interested in learning more about organ donation the mean percentage was 22.8% (range 0 – 76%, median 19%, standard deviation 17.3%, 49 respondents). ALS neurologists were queried on the average number of new ALS patients seen in their clinic annually, and the mean was 85.4 patients (range 5 – 400, median 72.5, standard deviation 72.5, 52 respondents).

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Discussion

Living organ donation is the ultimate way for a person to consciously give the "gift of life." Despite the tremendous need, this option is only available to healthy individuals and is currently denied to patients with terminal diseases, including ALS patients, despite their desire to participate. One such ALS patient in the United States said *"there's nothing greater than for a family member to receive an organ so they can watch their family grow up……..I am not suicidal, I just know that it is a matter of time before I die and wish to do a good thing for those people who have a good life expectancy."*[9] Such an altruistic desire has been expressed by many patients suffering from ALS, and has gone unfulfilled. Living organ donation is the most feasible option for these good Samaritans, as ALS does not lead to brain death, which provides maximum organ donation potential. The manner of death in ALS is respiratory failure and then circulatory arrest with subsequent poor organ perfusion. Organ donation in circulatory death is only possible after tracheostomy, ventilation and then withdrawal of life support to control the time of death. This is a high price to pay for most ALS patients, though it has happened at least once, where the patient said *"I am glad that in spite of my disease, there is still something I can do to help others in a significant way. ALS is preventing me from accomplishing what I wanted to do in my life, but hopefully, my donation will give others a chance to live out their dreams."*[13]

Our results indicate that a small portion of ALS neurologists routinely discuss living organ donation with their patients (5.9%), almost half report being asked by their ALS patients about this option (49%), and the majority do not have concerns with living organ donation in ALS patients (67.3%). Furthermore the majority of concerns expressed by the neurologists can be readily addressed.

Neurologist respondents expressed concern with the ALS donors risk incurred during organ recovery surgery and beyond (Figure 2). The highest concern is the risk of post-operative respiratory failure (PRF), followed by exposure to anesthetic agents, and renal dysfunction/ uropathy (due to single remaining kidney).

There are concerns that ALS patients without overt symptoms of respiratory failure may have subclinical diaphragmatic weakness, reduced respiratory reserve and thus be prone to PRF. However, this concern can be addressed by using the established guidelines for placement of percutaneous endoscopic gastrostomy (PEG) tubes in ALS patients, which are a FVC> 50% [14]. Additionally, there are multiple validated predictive scores (e.g. the ARISCAT Score) for the general population[15–17], which can be coupled with pulmonary function testing to provide an accurate risk assessment of PRF, enabling informed decisions by ALS patients.

A number of respondents expressed concern about worsening ALS symptoms after exposure to anesthetic agents and the systemic stress of surgery. These concerns are alleviated by abundant reports of anesthesia in a variety of surgeries in ALS patients, without evidence of decline. Surgeries have included feeding tube placement, diaphragmatic pacemaker implantation, spinal stem cell transplantation, and various other procedures[18,19,20,21].

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Concern for renal dysfunction or uropathy after kidney donation is an unlikely issue as there are multiple reports that living kidney donors have an extremely low peri-operative risk[22], and no increased risk of end stage renal disease (ESRD) when compared to the general population [23–26]. However, when compared to appropriate controls (i.e. those that fulfill kidney donation criteria) 2 studies show ESRD risk is slightly higher (0.27%) in living kidney donors[27,28]. The median time to develop ESRD is 8.6 years[27], which is significantly longer than the median survival of ALS patients (3 years)[10,11]. Furthermore, individualized risk projection tools for ESRD in living donor candidates are available[29]. With the available ESRD risk data, delayed progression data and risk projection tools, potential ALS living organ donors will be able to make an informed decision regarding their donation options and inherent risks.

The other area of concern for this practice centers on the possibility of disease transmission through an undetermined vector. The highest source of concern identified in this study was with a misfolded protein vector, followed by viral and gene transmission, which reflects possible factors of disease etiology [30]. In the unlikely event that a transmissible etiology is found for ALS, transplantation of a non-nervous system organ would be less likely to induce ALS in the recipient. There is ample experience with organ recovery and transplantation in patients with chronic hepatitis, and ALS donation could mirror this practice[31]. Transplants of ALS organs could also be limited to elderly patients (who may otherwise have a lower chance to be organ recipients), and significantly improve their quality of life.

It is evident from this survey that many ALS patients have spontaneously expressed interest and that there is a larger number that would likely consider living organ donation (1 in 4 patients). Yet, ALS centers are currently unable to honor such requests. There are no established mechanisms for screening, counseling or referral of these patients to transplant centers. With a reported mean of 85 new ALS patients per center per year, and 120 centers in the U.S., there is a potential of 2,550 ALS patients every year who wish to explore organ donation options [32]. These options are best considered when ALS patients have independent respiratory function (!>50% predicted) and are not severely debilitated. Current practice does not expose the donor to any financial costs or liability for the organ recovery surgery or post-recovery care. Honoring these wishes will provide ALS patients with a sense of altruism, selflessness and philanthropy, which will be of significant psychological benefit and provide them comfort at the end of life.

The medical concerns for living organ donation in this population can be readily addressed to ensure the ALS donor has adequate information to make an informed decision. Patients with ALS are a vulnerable population, which poses ethical concerns regarding living organ donation. These concerns need to be addressed with protective mechanisms such as formal ethics review, consultation with multiple physicians and specialties, psychological evaluation and support, psychiatric evaluation and depression screening, as well as a mandatory time period to consider the decision in every ALS patient who volunteers. Once these ethical concerns are addressed, Good Samaritan donation by ALS patients will not only increase the number of life-saving kidney transplants, but also ensure better quality of life in the recipients as compared to kidneys from deceased donors[33].

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To enable living organ donation in terminally ill patients, federal agencies and organ donation governing bodies should consider a *"donate life at the end of life"* program. This would establish processes, benchmarks and awareness for organ donation in this population, and remove the penalties incurred by transplant centers and surgeons when terminally ill donors suffer clinical morbidity or mortality related to their primary disease and unrelated to organ recovery surgery[6].

Our study has several limitations inherent to survey-based studies in general, such as small sample size, recall, belief and reporting bias. Survey respondents are also assumed to have different depths of knowledge regarding organ donation. This is a survey of ALS neurologists and not patients, and therefore, subject to neurologist's perceptions and biases. However, the strengths of this study are its high response rate, of 42.9% which is robust compared to the reported literature[34]. Our respondents were well distributed geographically across the United States of America (figure 1). Our questionnaire though comprehensive, was simple and easy to administer and revealed meaningful findings. This study is being followed with a suitably designed survey administered directly to ALS patients, to assess their interest and concerns with living organ donation and "donating life at the end of life".

Supplementary Material

Refer to Web version on PubMed Central for supplementary material.

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Highlights

- ALS patients are currently unable to become living organ donors due to current policies with donation.
- Transplant centers are penalized if a living donor suffers morbidity even if due to terminal illness and not donation.
- Our survey indicates that majority of neurologists (67%) do not have any concerns with ALS living organ donation.
- Medical concerns expressed by ALS neurologists can be readily addressed in a systematic way.
- Living organ donation in ALS patients needs to be explored to provide this valuable altruistic option to these patients.

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Figure 1. Geographical distribution of ALS neurologists surveyed in the United States

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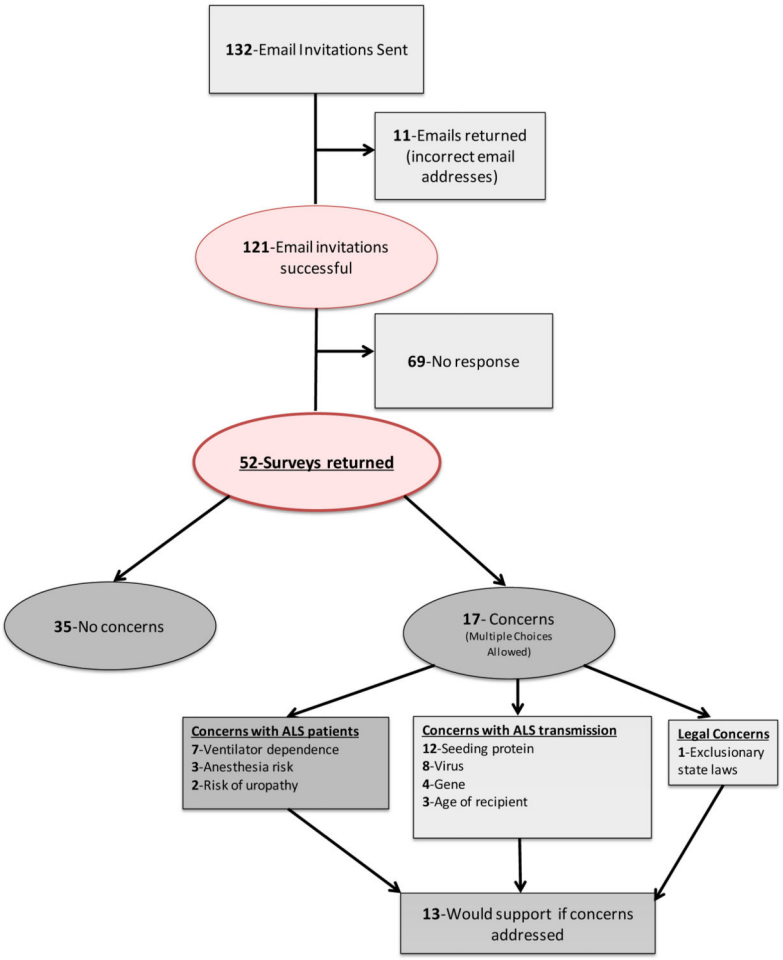

Figure 2.
Flowchart depicting survey response rates and results.