

Grant Application to the Blazeman Foundation for ALS

Title: Establishment of the Blazeman ALS Tissue Collection

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Background:

Amyotrophic Lateral Sclerosis (ALS) is a devastating motor neuron disorder that traps a fully cognitively functioning person in a body unable to initiate or control voluntary movements. A small percentage of individuals with ALS have an inherited component (5-10%) due to a deficiency of the enzyme superoxide dismutase (SOD1). However, the underlying mechanism in the majority of individuals with ALS has not yet been identified. Currently there is no cure for ALS, nor has there been any significant progress in drug development.

Research to find the cause and underlying mechanism is occurring at several levels. The majority of studies involve living patients. However, certain studies that address the pathology and biochemical basis of ALS can only be studied by looking directly at the tissue involved. For ALS this requires study of the post-mortem spinal cord. The Blazeman Foundation has already made a very personal donation by donating brain and spinal cord tissue from Jonathan Blais in 2007.

Tissue donation can only proceed with the consent of the next of kin after an ALS patient dies. Two aspects have to be considered: 1) have ALS patients and their families been counseled about the feasibility of tissue donation for research and 2) is there a mechanism in place to collect, store and distribute tissue? It is the experience of the NICHD Brain and Tissue Bank that cognitively active adult with a terminal illness are favorably inclined to consider tissue donation. Unfortunately, medical personnel often do not discuss the possibility of tissue donation with ALS patients or their families out of a misplaced sensitivity of mentioning the issue. Therefore an outreach program and publicity regarding post-mortem tissue donation is necessary. This activity is best handled by support groups that can speak with ALS families because their only motive is the well being of ALS families. Currently there is no centralized collection center for ALS tissue in the United States. The few centers that collect ALS tissue have limited resources for storage, advertisement of tissue availability, distribution of tissue, and mechanism for review of tissue request.

Study of ALS requires a comparison of ALS tissue with normal tissue. Therefore, donation of post-mortem tissue from unaffected family members is gratefully accepted. Tissue from family members may also provide information if specific genes are missing or activated in the ALS individual.

Tissue donation is recognized as a magnanimous act by all major religions. However, the decision to donate is a very personal one. No one answer is appropriate for all. But for those individuals and families that make this decision to donate tissue, a procedure is available by which you can leave a legacy for ALS research. There is no cost to the family as the Blazeman Foundation, through its funding of this grant, and the National Institute of Health covers all costs related to tissue recovery. It does not delay the funeral because the tissue has to be recovered within 24 hours since the quality of the tissue, and therefore its potential for important ALS research, deteriorates with time after death. Tissue donation is compatible with an open viewing.

The NICHD Brain and Tissue Bank has a wide mandate to collect tissue, including from ALS donors. The NICHD Bank has been funded since 1991 and has collected tissue from over 3000 cases in every continental state of the US as well as in several foreign countries. The tissue is stored in modern facilities constructed for the Bank by the University of Maryland. Safety features include emergency electrical power back up, liquid carbon dioxide back up for each freezer, and electronic monitoring system of freezer status. Over 800 researchers in 21 countries have received approximately 30,000 tissue samples. The researchers have published 500 papers based on research performed with tissue received from the Bank. Specifically, the Bank has already supported ALS research. Seventeen ALS patients and their families have donated spinal cord and brain tissue to the Bank. A critical advantage of the NICHD Bank is that the Bank has normal tissue that is vital for comparison purposes.

However, to accelerate ALS research more tissue is required as well as financial support for researchers. Through support of the Blazeman Foundation more individuals,

especially athletes with ALS, will decide to donate tissue to the Bank. Funds from the Blazeman Foundation will pay for a significant part of the cost of making a successful donation. Cost for the recovery of the brain and spinal cord ranges between \$1,000 and \$1,800. Transport of the donor to and from the collection site may cost \$200 - \$500. A neuropathology report is required for the proper diagnosis of ALS. The cost of preparation of slides and evaluation by a neuropathologist ranges from \$500 to \$800. It is estimated that direct costs for collection of tissue from one donor will range from \$1,700 to \$3,100. This does not include the service of the NICHD Brain and Tissue Bank staff involved in tissue recovery, processing and storage. The latter costs will part of the overall operating expenses of the NICHD Brain and Tissue Bank. For calculating purposes an average of \$2,000 per case will be used to calculate the recovery cost of donated tissue from one ALS donor. Funds received from the Blazeman Foundation will significantly increase the availability of ALS tissue for research.

Tissue to be collected: spinal cord, brain, peripheral nerve and CSF. A case will not be collected if it is not possible to collect the spinal cord since the diagnosis of ALS cannot be made without examining the spinal cord for its distinctive neurodegeneration. Recovery of the spinal cord may not be feasible in cases where a pathologist is not able or willing to remove the spinal cord due to time, availability of facility, or other reasons.

Reporting:

A synopsis of activities will be submitted on a yearly basis.

Budget:

The grant will be for a 2 year period. Either side has the right to terminate the project. The Blazeman Foundation may also increase the funds for the grant at their discretion. Funds may be used for travel for ALS related activities only after prior consultation and approval by the Blazeman Foundation.