

THE MIKEY CZECH FOUNDATION-FUNDED PHASE II CLINICAL TRIAL YIELDS PEDIATRIC BRAIN TUMOR BREAKTHROUGH AT HARVARD MEDICAL SCHOOL - DANA-FARBER CANCER INSTITUTE

New Canaan, CT – October 23, 2014 – The Mikey Czech Foundation, Inc. (the “Foundation”, www.mikeyczech.org) a 501 (c)(3) non-profit foundation established by the Stephen J. Czech family to finance medical research for pediatric brain tumors, announced a major breakthrough related to the discovery of mutations that cause Diffuse Intrinsic Pontine Glioma (“DIPG”) pediatric brain tumors, the type of tumor that claimed the life of the Czech’s only son, James Michael “Mikey” Czech, in 2008 at age 11.

DIPG brain stem tumors affect the cranial nerves of children, ages 3 – 16 years old, destroying the nerves that supply the muscles of the eye and face, and muscles involved in swallowing. Symptoms include double vision, inability to close the eyelids completely, “drooping” on one side of the face, and difficulty chewing and swallowing. The tumor also affects the “long tracks” of the brain, with resultant weakness of the arms or legs and difficulty with speech and walking. As the tumor grows, children initially lose their ability to use their limbs. Thereafter, they sequentially lose control of their bowels, hearing, vision and their ability to swallow and breathe. Notwithstanding their physical deterioration, the children maintain their mental faculties, are fully cognizant of what is happening to them, and experience pain comparable to that of being skinned alive. Survival past 9 to 12 months is uncommon, and new approaches to treating these tumors are urgently needed as no remedies currently exist to treat DIPGs.

“The assets required to cure DIPGs consist of: (a) a sustainable funding source; (b) world-class neuro-oncologists; and (c) state-of-the-art research facilities,” said Steve Czech, Mikey’s father and Chairman & Co-Founder of The Mikey Czech Foundation. “We intend to be the sustainable funding source and we believe that Dr. Kieran, his colleagues and Harvard Medical School/Dana-Farber represent the “best-in-class” with respect to pediatric neuro-oncology research and state-of-the-art research facilities.

In addition to his efforts with the Foundation, Czech is the Managing Partner and Chief Investment Officer of Czech Asset Management, L.P., a credit focused alternative investment manager based in Greenwich, CT. In an effort to establish a sustainable funding source for DIPG research, Czech contributes a percentage of his personal investment profits each year to the Foundation and DIPG research.

WHAT IS THE OBJECTIVE OF THE CLINICAL TRIAL BEING FUNDED BY THE FOUNDATION?

- The Mikey Czech Foundation is currently funding a Phase II clinical trial at Harvard Medical School/Dana-Farber Cancer Institute in Boston, MA.
- The objective of the trial: “*A Phase II Trial of Molecularly Determined Treatment of Children and Young Adults with Newly Diagnosed Diffuse Intrinsic Pontine Glioma*” is to use genomic knowledge to develop effective therapies for pediatric DIPGs.

WHO IS CONDUCTING THE CLINICAL TRIAL?

- The trial is led by Dr. Mark W. Kieran, MD. PhD, Clinical Director of the Pediatric Brain Tumor Center at Harvard Medical School/Dana-Farber Cancer Institute.
- He has developed an international reputation as a leader in pediatric neuro-oncology.
- Dr. Kieran is currently the lead investigator on more than 10 clinical trials through national consortium, investigator initiated or pharmaceutical sponsored clinical trials.
- He has a number of ongoing Material Transfer Agreements (“MTAs”) with pharmaceutical and biotechnology companies to support early translation of novel drugs.
- Kieran also holds six (6) Investigational New Drug applications (“INDs”) for experimental therapies in children.

- Additionally, Dr. Kieran is co-chair of the malignant glioma committee of COG, and on advisory and review committees of numerous national foundations as well as the Food and Drug Administration (“FDA”).

WHAT INSTITUTIONS ARE PARTICIPATING IN THE CLINICAL TRIAL?

- UT Southwestern Medical Center
- Ann & Robert H. Lurie Children’s Hospital of Chicago
- Cook Children’s Medical Center Fort Worth, TX
- Doernbecher Children’s Hospital Oregon
- Johns Hopkins Medical Center
- Penn State Hershey Medical Center
- Seattle Children’s Hospital
- Washington University St. Louis
- Children’s Hospitals and Clinics of Minnesota
- Children’s Hospital Colorado
- New York University Medical Center
- University of California San Francisco
- University of Louisville/Kosair Children’s Hospital
- Children’s Hospital Los Angeles Children’s Healthcare of Atlanta
- Miami Children’s Hospital
- Children’s Hospital of Michigan
- Duke University Medical Center

WHAT ARE THE RESULTS OF THE CLINICAL TRIAL TO-DATE?

FOR THE FIRST TIME EVER...

- Dr. Kieran was able to biopsy live DIPG tumor tissue.
- Using the latest advances in nano-technology and biomedical engineering techniques and in conjunction with The Broad Institute at M.I.T., Dr. Kieran sequenced 40 tumor biopsy samples, of which 25 were from patients with DIPG tumors.

FOR THE FIRST TIME EVER...

- The sequencing efforts identified a number of errors (“Mutations”) in the DNA code and epigenetic changes, which alter how genes are expressed.
- At least two of the new mutations in the FGFR1 and PI3K cell growth-signaling pathways might be susceptible to blocking by *existing* drugs that are being tested in other cancers.
- The other two molecular targets that they uncovered – including mutations in the H3F3A protein and the gene ACVR1 – lack specific drugs, but provide new opportunities for future treatment development.

FOR THE FIRST TIME EVER...

- The ACVR1 mutation was found in five of the DIPGs and it is the *first time EVER that this gene has been implicated in cancer!*

THE BOTTOM LINE

- As a result of the efforts of The Mikey Czech Foundation, there has been more progress made in understanding the biology of DIPG tumors in the last three years than there has been in the prior 50 years!
- Based on the results of the aforementioned clinical trial discoveries, we now have a better idea of the underlying mutations that lead to the development of this terrible disease.
- Most important, these mutations can now be targeted, the first step in identifying drugs that will be required to effectively treat these tumors.

About DIPG. DIPG brain stem tumors affect the cranial nerves of children, ages 3 – 16 years old, destroying the nerves that supply the muscles of the eye and face, and muscles involved in swallowing. Symptoms include double vision, inability to close the eyelids completely, "drooping" on one side of the face, and difficulty chewing and swallowing. The tumor also affects the "long tracks" of the brain, with resultant weakness of the arms or legs and difficulty with speech and walking. As the tumor grows, children initially lose their ability to use their limbs. Thereafter, they sequentially lose control of their bowels, hearing, vision and their ability to swallow and breathe. Notwithstanding their physical deterioration, the children maintain their mental faculties, are fully cognizant of what is happening to them, and experience pain comparable to that of being skinned alive. Survival past 9 to 12 months is uncommon, and new approaches to treating these tumors are urgently needed as no remedies currently exist to treat DIPGs.

About Dana-Farber Cancer Institute. Dana-Farber Cancer Institute is a principal teaching affiliate of the Harvard Medical School and is among the leading cancer research and care centers in the United States. It is a founding member of the Dana-Farber/Harvard Cancer Center (DF/HCC), designated a comprehensive cancer center by the National Cancer Institute. It provides adult cancer care with Brigham and Women's Hospital as Dana-Farber/Brigham and Women's Cancer Center and it provides pediatric care with Boston Children's Hospital as Dana-Farber/Children's Hospital Cancer Center. Dana-Farber is the top ranked cancer center in New England, according to *U.S. News & World Report*, and one of the largest recipients among independent hospitals of National Cancer Institute and National Institutes of Health grant funding. Follow Dana-Farber on Twitter: [@danafarber](#) or [Facebook](#).

About Czech Asset Management, L.P. Czech Asset Management, L.P. is a Greenwich, CT-based direct lending firm engaged in the business of originating and investing in asset-based and cash-flow first and second lien secured floating rate loans for middle market borrowers located throughout North America and Europe. www.czechamlp.com.

Contact:

Stephen J. Czech
Chairman & Co-Founder
The Mikey Czech Foundation,
Inc. P.O. Box 1717
New Canaan, CT 06840
Phone: 203-769-8401
Email: sczech@sjccapital.com