

Model Systems Development

creating tools needed to study chordoma

Why are Models Important?

Model systems are research tools that mimic the behavior of human tumors, and allow researchers to study cancer without experimenting on people. They are needed to develop virtually any type of new therapy, however limited availability of valid models of chordoma is preventing progress from being made. Therefore, developing new models is urgently needed to advance research and improve the treatment options available to chordoma patients.

There are three main types of models that scientists use to study cancer: cell lines, xenographs, and transgenic organisms. Each of these models serves an important purpose, and has its own unique benefits and drawbacks. Using all three models in combination gives scientists the most complete understanding of how the disease works and how to treat it.

- » **Cell Lines** are live tumor cells grown perpetually in artificial vessels like Petri dishes. They are the easiest of any model to grow and manipulate, and are therefore the tool of choice to answer most questions about chordoma biology, and to test large numbers of drugs.
- » **Xenographs** are human tumors that are implanted and grown inside laboratory animals. Like cell lines, they are used to test the response of the tumors to various drugs. The primary advantage of xenografts is that they are grown inside a living organism under conditions that more closely mimic actual human tumors.
- » **Transgenic organisms** are genetically engineered to develop tumors that mimic specific human cancers. Transgenic mouse models are particularly valuable for understanding the underlying biology of human tumors, and like xenografts, may also be useful for screening new drugs.

Chordoma Foundation Approach

There are currently only two valid chordoma cell lines in the world, and no xenographs or transgenic models exist yet. Until these vital models are available little can be done to develop and test new treatments for chordoma. Therefore, the Chordoma Foundation is focused on developing, studying, and distributing valid models through three complementary programs.

» **Model Systems Grant Program**

The Chordoma Foundation provides one-year grants (~\$25,000) to create and characterize new models of chordoma. In 2008 Model Systems Grants were awarded to researchers at Duke University, Harvard, Istituto di Tumori Milano, Johns Hopkins University, University College London, and University of Florida.

» **Cell Line Panel**

Some chordoma cell lines reported in the literature have turned out not to be chordoma (some were not even human). Studying invalid cell lines produces invalid data and is an enormous waste of time and money. Therefore the Chordoma Foundation is creating a panel of 15-20 well-characterized and validated chordoma cell lines, each of which faithfully represents the tumor from which it was derived. Data generated on each cell line will be aggregated in a centralized database, allowing results from the same cell line in multiple labs to be directly compared. By enabling all researchers to study the same set of high-quality cell lines, the Cell Line Panel will make the entire field of chordoma research more efficient and effective, and will accelerate the pace of discovery.

» **Cell Line Prize**

In order to spur development of new cell lines in as many labs as possible the Chordoma Foundation will award a \$10,000 unrestricted prize for each validated cell line that is accepted into the Chordoma Foundation Cell Line Panel. Already one prize has been awarded to the University of Ulm in Germany for creating the second chordoma cell line.