

Clear Cell Sarcoma Project Update

July/August 2019



CCS Nanocourse at cc-TDI

The annual cc-TDI nanocourse was held from August 12 – 16, 2019. Our Scientific Director, Dr. Charles Keller, presented on cc-TDI's latest efforts on developing CCS-focused therapeutics. Our guest speaker list included:

- Agnieszka Wozniak, PhD (KU Leuven) – CCS Clinical Trials
- Jeff Toretsky, MD (Georgetown) – Ewings Family of Tumors Overview
- Paul Huang, PhD (ICR London) – Sarcoma Proteomics
- Kevin Jones, MD (University of Utah) – Mouse Models of CCS
- Tyuji Hoshino, PhD (Chiba University) – Drug Design
- Gwenn Hansen, PhD and Steve Basham, PhD (Nurix Pharmaceuticals) – Programs in Proteome Therapeutics
- Xiangshu Xiao, PhD (OHSU) – Development of 666-15
- Michael Cohen, PhD (OHSU) – EWSR1 Drug Development
- Viktors Berstis (WorldCommunityGrid.org) – *Nanocourse Plenary Speaker*
- Ganapati Srinivasa (ODA) – Genomics
- Tommy Pham (Nike) – Industry Partnerships

Please see the below links for additional CCS talks from the 2018 and 2019 Nanocourses:

Nanocourse 2018 - CCS:

<https://childrenscancertherapydeve.box.com/s/7o66ufaoe55gu4dw6shyxs6lwrk9pk9b>

Nanocourse 2019 - CCS:

<https://childrenscancertherapydeve.box.com/s/av0wl1jcotxpk4e3ipv8m7w3pesrigo4>

Our special thanks to Sara's Cure for their contributions to helping make the 2019 Nanocourse a success!

Collection of CCS Tumor Resources at cc-TDI

One of our goals at cc-TDI has been to create a centralized biobank of all available CCS resources. Thus far, we have collected numerous specimens including 8 cells lines (from 6 different patients) and tissue specimens from 10 different patients. Our list of resources is shown in Table 1. We continue to accrue additional resources as they become available.

Table 1. Collections of CCS Tumor Resources at cc-TDI

	same patient								same patient										
	<i>SU-CCS-1</i>	<i>K4S</i>	<i>CCS281</i>	<i>IMP-CCS-SY</i>	<i>NCC-CCS/B-X2-C1</i>	<i>NCC-CCS/B-C1</i>	<i>NCC-CCS1-X4-C1</i>	<i>RSAR001</i>	<i>CF-00465-1</i>	<i>CF-00732</i>	<i>CF-00733</i>	<i>CF-00734</i>	<i>CF-00464-1</i>	<i>CF-00464-2</i>	<i>CF-00466</i>	<i>CF-00481</i>	<i>CF-00486</i>	<i>CF-00491</i>	<i>CF-00493</i>
Age	16	25	unk	17	43	43	43	43	28	63	15	21	17	19	22	18	13	25	49
Gender	F	M	unk	F	F	F	F	M	M	F	M	M	F	F	F	F	M	F	F
Location	Heel	Node	unk	Ankle	Leg	Leg	Leg	Pleural fluid	GI	GI met	GI	GI	GI	GI met	GI met	GI	Axillary	Leg	Foot
DNA Sequenced	✓	✓	✓	✓	✓	✓	✓	✓	✓			✓	✓	✓	✓	✓			
RNA Sequenced	✓	✓	✓	✓	✓	✓	✓	✓	✓		✓	✓	✓	✓	✓	✓			
V5 Drug Screened	✓	✓	✓	✓	✓	✓	✓	✓											

Immortal Cell Line
 FFPE Tissue

Drug Screening for CCS

A group of 60+ compounds have been chosen for testing with all of the CCS cell lines available at cc-TDI. These agents were selected based on a review of the current literature as well as consultation with multiple pediatric and young adult sarcoma oncologists for the most promising therapeutics in various stages of development. Presently, all 8 CCS cell lines shown in Table 1 have run through this drug screening process. Two additional cell lines (normal human kidney cells, HEK293, and normal mouse fibroblasts, C2C12) were included as negative controls. We have also tested two positive control cell lines for the EWSR1 translocation (Ewings sarcoma cell lines A-673 and TC71). Results of this study are currently under review and a manuscript is currently in preparation for public dissemination.

Fundraising

A special thanks to Sara's Cure for all of their continued efforts dedicated to funding CCS-focused research at cc-TDI!

CureFAST Program at cc-TDI

This is just a reminder that we are actively enrolling patients into our CuRe-FAST Program, whereby tumor samples can be donated to cc-TDI and used for research purposes, free of charge. Please contact Andy Woods (andy@cc-tdi) for more information and to enroll.