HCI Named a Von Hippel Lindau Clinical Care Center

In December 2016, the VHL Alliance (VHLA) named Huntsman Cancer Institute (HCI) at the University of Utah a VHL clinical care center. The VHLA is a nonprofit organization that supports research and education about Von Hippel Lindau Syndrome (VHL).

VHL raises the risk for some cancers and other tumors. The tumors can occur in the brain, spine, kidneys, or other organs. The syndrome affects about 200,000 people worldwide.

The VHL Clinical Care Clinic at HCI lets VHL patients get all screening and treatment in one place. Led by a genetic counselor and genetics nurse, HCI’s specialists work together with patients to plan the best course of treatment. The team includes imaging specialists, surgeons, and other doctors. The clinic also helps patients find and participate in research studies and clinical trials.

The clinic will also serve patients with other conditions that have similar types of tumor risk. These include Birt-Hogg-Dube (BHD) and hereditary leiomyomatosis and renal cell cancer (HLRCC).
Meet the First Patient of HCI’s Von Hippel Lindau Comprehensive Clinic

Mikaela is the first patient of the Von Hippel Lindau (VHL) Comprehensive Care Clinic at Huntsman Cancer Institute (HCI). Doctors diagnosed the 18-year-old with VHL several months after she had surgery to remove tumors in her brain, neck, and spine.

“I’d never heard of VHL before,” Mikaela says. “It took many doctor visits for me to understand the disease. One of the hardest parts was trying to find doctors who even know what it was.”

One of her doctors referred Mikaela to HCI’s VHL Clinic. “I was a little skeptical, considering that I don’t have cancer,” she says. “Our first appointment lifted a huge weight from my shoulders. The VHL team is kind and informative. And they seem to know everything there is to know about VHL.”

Mikaela loves being active. She grew up camping, hiking, fishing, and “playing every sport out there.” She says her main concerns about VHL had to do with her quality of life. Even after multiple surgeries, tumors and cysts still remain.

“I have daily pain and struggles related to this disease, but it isn’t stopping me,” Mikaela says. “I play on my high school’s varsity softball team, attend school daily, and go out with my friends whenever possible. I am not my disease…I am still a normal teenager.”

Like Mikaela, most patients already know they have VHL at their first visit to the clinic. “We work with them to set up a schedule for follow-up care,” says Samantha Greenberg, LCGC, an HCI genetic counselor who oversees the VHL clinic. “Patients without current symptoms usually have annual follow-ups.”

Greenberg says most adults with VHL need to see four to six different doctors each year. Keeping track of an appointment schedule this complex can be a difficult task. “HCI’s VHL Comprehensive Care Clinic lets patients see all the specialists necessary at a central location,” she says. “It’s not always possible, but we try to schedule all their appointments for the same day.”

Mikaela’s advice to others who have VHL is clear: “Do not let your disease keep you from doing what you love. Do not let it hold you back from living the life you want to live.”
The FAP erlotinib and sulindac trial (FAPEST) tested whether the combination of the two FDA-approved medicines can stop small bowel (duodenal) polyps from growing. Huntsman Cancer Institute (HCI) investigators Randall Burt, MD; Jewel Samadder, MD; and Deb Neklason, PhD, led the clinical trial. The study was published in *JAMA* in 2016.

Study participants all had familial adenomatous polyposis (FAP) or attenuated FAP. FAP patients develop precancerous polyps in the colon and the small bowel. The polyps must be removed before they become cancer.

Erlotinib is a chemotherapy drug used to treat some types of cancer. Sulindac is a nonsteroidal anti-inflammatory drug (NSAID) similar to aspirin. Half the patients in the study received the drug combination. The other half received a pill that looked identical without the drugs (placebo). The study compared the number and size of duodenal polyps in the two patient groups.

After 6 months of treatment, patients who took the drug combination had an overall decrease in the number of polyps. Patients who took the placebo had an overall increase in the number of polyps.

The study showed that the drug combination helps slow and decrease duodenal polyp growth. “The positive effect of this drug combination was so clear that we closed the study earlier than planned,” says Neklason.

Many study participants had uncomfortable side effects. The most common was a skin rash. More research needs to be done before these drugs can be regularly prescribed. This could include studies to find dosages that will reduce polyps with fewer side effects. Other studies could test how well the drug combination works over the long term.

Annual exams with a specialist are the most important way to monitor and manage FAP. Each person needs his or her own specialized disease management plan that may differ from those of other family members.

To find clinical trials in your area, visit [clinicaltrials.gov](http://clinicaltrials.gov).

For patient-friendly information about hereditary colon cancer conditions, visit [www.hcctakesguts.org](http://www.hcctakesguts.org).
A complete family medical history is one of the most important tools your health care team uses to manage cancer risk for you and your family. When you collect, record, and share this information, you leave a legacy for your family now and for generations to come.

Talk with your close family members—parents, brothers, sisters, aunts, and uncles. Ask them to talk about their brothers, sisters, parents, aunts, uncles, and grandparents. These questions will help you get the family history information health care teams use:

- Did [this relative] have any health issues? What were they?
- Was [this relative] ever diagnosed with cancer? What type of cancer? How old was [this relative] when diagnosed?
- What is our ancestry? What part of the world did our family come from?

You may find it hard to bring up topics such as cancer and relatives who have passed on. It is worth the effort. Sharing this information can benefit your family’s health.

Record the information in a Google document or other shareable resource so the whole family can use it with their health care teams. Family members can also update the shared document as needed.

Reunions, holiday dinners, and other family gatherings are good times to start a conversation about family medical history. But you can start anytime—call, write letters, or send e-mails.

You can search the Utah Department of Health website, http://health.utah.gov, for their Family Health History Toolkit.

Your kids can help, too! Find a family medical history activity plan at http://teach.genetics.utah.edu.

Scroll to the alphabetical list and choose “Family Health History.” From there, look for the unit called “What’s Your Family Health Story?”