



## CRAVAT Foundation

# What are Vascular Tumors?

The **C**enter for **R**esearch and **A**nalysis of **V**ascular **T**umors (CRAVAT) is raising awareness of and harnessing research to address one of the rarest forms of cancer, vascular tumors. CRAVAT has its origins in a small but effective support group called HEARD (**H**emangioendothelioma, **E**pithelioid Hemangioendothelioma And Related Disorders),

### Definitions:

Vascular cancers (ie, cancers pertaining to vessels, particularly blood vessels) are very rare, accounting for only a fraction of 1% of all cancers. Hemangioendotheliomas are a diverse group of vascular cancers. The term is descriptive as the predominant lesional cell is a precursor of an endothelial cell. Hemangioendothelioma is distinct, both histologically and pathologically, from the common hemangiomas of infancy. It is considered a low grade malignancy and can involve soft tissue, bone, skin, liver, lymph node and/or lung.

Hemangioendotheliomas and hemangiopericytomas as a group have a malignant potential between benign hemangiomas and highly malignant angiosarcomas. Hemangioendotheliomas do not respond to either standard chemotherapy or radiation. Patients' outcomes are unpredictable as these tumors often have extended periods of quiescence punctuated by rapid growth. Liver transplant is offered to patients with liver tumors but there is no currently accepted medical (non-surgical) treatment for this group of diseases.

Both hemangiomas and angiosarcomas are more common than hemangioendotheliomas and therefore receive far more scientific attention and funding. CRAVAT was formed to rectify this disparity and assure that hemangioendotheliomas are no longer forgotten or ignored as described above by Cynthia.

## Subgroups

There are clear patterns of clinical presentation and specific subgroups of hemangioendothelioma. For example Kaposiform Hemangioendothelioma (KHE) typically occurs on the trunk, remains very local and is commonly associated with coagulopathy (abnormal blood clotting). In contrast, epithelioid hemangioendothelioma (EHE) is most common in the liver, commonly occurs at multiple sites at the same time (metachronous growth), and does not cause coagulopathy.

The pattern of progression of these tumors varies greatly and is very different from its benign counterpart, the hemangioma. While in rare cases spontaneous regressions have been reported, these tumors are generally slow and indolent. However, in some cases it can progress to devastating disease. The tumors are often locally invasive and destroy surrounding tissue.

Other forms include the benign infantile hemangioendothelioma which generally regresses within the first few years of life; and depending on the histologic appearance of the lesional cell, a variety of even rarer subtypes of hemangioendothelioma, including: spindle cell, retiform, composite, papillary and polymorphous.

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