The VHL Alliance (VHLA, [vhl.org](http://www.vhl.org)) is dedicated to research, education, and patient support to improve awareness, diagnosis, treatment, and quality of life for those affected by VHL. We very much appreciate your interest in the VHL Clinical Care Center Program; your participation will help us achieve this mission.

VHLA recognized Clinical Care Centers (CCCs) are committed to providing outstanding holistic, coordinated care for VHL patients. All applications are reviewed by our Clinical Advisory Council ([www.vhl.org/ about/people/clinical-advisory-council/](file:///C:\Users\isussman.VHL\Dropbox%20(VHL)\Website%20Restructure%202015\PDF%20Templates\www.vhl.org\%20about\people\clinical-advisory-council\)). Acceptance into the program is not automatic, and patient feedback is always carefully considered. It is not a requirement to have every specialist outlined under one roof, but close proximity is useful. VHLA has two categories of care centers: Comprehensive Clinical Care Center (CCCC) and Clinical Care Center. Each center is further designated as Adult, Pediatric, or All Ages. Specialists who are required to be involved at every recognized center are indicated by an “**\*”** and highlighted in yellow**.** Additional specialties required only for CCCC’s are denoted by “\*\*” and highlighted in blue. Please note if you plan to use the services of another physician at a sister institution.

Institutions will be designated as a CCCC based on 1) this application and 2) patient feedback, solicited by VHLA, on center performance. In order to evaluate patient feedback, no new center will be approved as a Comprehensive until the center has functioned as a VHL Clinical Care Center for a minimum of 2 years.

VHLA is working with the HLRCC Family Alliance (Hereditary Leiomyomatosis and Renal Cell Cancer ([www.hlrccinfo.org](http://www.hlrccinfo.org), at which a HLRCC Handbook and surveillance guidelines can be found) and BHD (Birt-Hogg-Dubé, [www.bhdsyndrome.org](http://www.bhdsyndrome.org/)). As a result, we are expanding our Clinical Care Center program to include these groups of patients. As such, the specialties of Gynecology, Pulmonology, and Dermatology have been added to the list of clinical specialties.

VHLA encourages all Clinical Care Centers to provide educational opportunities for physicians and staff on the current clinical practices for VHL, HLRCC, and BHD. Such training opportunities would include attendance at the annual VHLA-sponsored patient educational meeting (VHLA Annual Family Meeting), the biennial International VHL Medical Symposium, or on-site lecture presentations by experts caring for VHL / HLRCC / BHD patients. In addition, each specialist included in an approved application will be added to the appropriate VHLA list serve and can participate in clinical care discussions with their colleagues.

When applying to be a Clinical Care Center, please provide as much of the following as possible and email as a Word file saved as *Submission Date YYYY.MM.DD – Abbreviated Institution Name– CCC Application,* (ie: 2019.01.30-UMich-CCC Application) to: [josh.mann@vhl.org](mailto:josh.mann@vhl.org)

On behalf of the VHL Alliance and for all involved patients and their families—we thank you for your interest in helping us achieve our mission!

Sincerely,

|  |  |  |
| --- | --- | --- |
| **Chandra Clark, MEd**  VHLA Executive Director | Description: OIsign-1  **Othon Iliopoulos, MD**  VHLA Clinical Advisory Council Chair, Research Council Chair, Board Member | **C:\Users\Ilene\Downloads\Joshua-Mann (1).jpg**  **Joshua Mann, MPH**  VHLA Director of Health |

Application Contents

[CCC Process Matrix 3](#_Toc38711568)

[CCC Requirements 3](#_Toc38711569)

[Responsibilities of Each CCC Team Member 5](#_Toc38711570)

[CCC / VHLA Liaison 5](#_Toc38711571)

[CCC Sponsoring Physician 5](#_Toc38711572)

[CCC Point of Contact 5](#_Toc38711573)

[CCC Patient Navigator 5](#_Toc38711574)

[CCC Specialist 5](#_Toc38711575)

[Application Form: VHL Clinical Care Center 6](#_Toc38711576)

[General Information 6](#_Toc38711577)

[CCC Lead Team Information 6](#_Toc38711578)

[CCC Specialists’ Information 6](#_Toc38711579)

[Services Provided 10](#_Toc38711580)

[Short Answer Questions 11](#_Toc38711581)

[Clinical Care Center Programmatic Opportunities 12](#_Toc38711582)

[Core Requirements 12](#_Toc38711583)

[Additional Programmatic Opportunities 12](#_Toc38711584)

[Suggested Psychosocial Questionnaire 13](#_Toc38711585)

[VHLA Suggested VHL Surveillance Guidelines 14](#_Toc38711586)

[Suggested HLRCC Surveillance Guidelines 17](#_Toc38711587)

[Suggested Referral Criteria to VHL Clinical Care Center 18](#_Toc38711588)

# CCC Process Matrix

Below is a process matrix that demonstrates how patients contact a CCC, how CCC team members work together to provide coordinated care, and how the Liaison works with the CCC. See next section for complete list of team member responsibilities.

VHL Navigator introduces Patient to Liaison

Patient contacts CCC via Point of Contact

Patient contacts CCC Specialist / Department

# CCC Requirements

**Each Clinical Care Center application (both new centers and renewals) are required to:**

* Submit institutional surveillance protocol to be approved by Clinical Advisory Council.
* Provide surveillance protocol, a list of participating team members and their contact information, and referral standards to the specified Point of Contact for that Center. (This will help them in the referral process as they help the patient navigate the system.)
* Identify and implement system to assess psychosocial needs of each patient. (Addressing ongoing psychosocial needs may be better achieved at a clinic closer to the patient’s home, but the patient navigator and referring specialists at the CCC should work with the patient / family to establish this care.)
* Agree to communicate medical updates, including psychosocial needs, with patient’s primary care clinician.
* Identified team members must be Board certified in specified area. Retinal specialists are required to have completed a retina fellowship.
* Agree to turn in complete, updated applications every other year to keep contacts current.
* Encourage patients to participate longitudinally in the MyVHL: Patient Natural History Study (vhl.org/MyVHL).

|  |  |  |  |  |
| --- | --- | --- | --- | --- |
|  | **Required** | | **Optional** | |
| Specialists | * Genetic Counselor * Ophthalmologist * Neurosurgeon * Urologist * Endocrinologist | * \*Nephrologist * \*Endocrine Surgeon * \*Neurootologist * \*Oncologist * \*Radiologist | * Geneticist * Neurology * Neurooncologist * Surgical Oncologist * Gynecologist | |
| * Pediatrician (if center sees children)   *\*Additional Required Specialists for CCCCs* | | * Pancreatic Gastroenterologist * Nutritionist (inpatient and outpatient) * Maternal-Fetal Medicine Specialist | |
| Surveillance | * MRI scanner * CT scanner * MIBG * Audiology Testing | * Genetic Testing * Eye / Retinal Exam * Fluorescein Angiography * Psychosocial Testing | * PET scanner * PET / CT scanner * Ultrasound | |
| Therapy |  | | * Physical Therapy * Occupational Therapy * Speech Therapy | * Neurological Rehabilitation * Oncology Support Groups |
| Treatment | * Stereotactic Radiosurgery * Radio Frequency Ablation * Cryoablation * Open Partial Nephrectomy * Laproscopic Partial Nephrectomy * Open Partial Adrenalectomy * Laproscopic Partial Adrenalectomy * Reoperative Partial Adrenalectomy * Retinal Laser Surgery * Retinal Cryosurgery | | * Robotic Partial Nephrectomy * Robotic Partial Adrenalectomy * Resection of Retroperitoneal Paragangliomas | |

# Responsibilities of Each CCC Team Member

## CCC / VHLA Liaison

* Volunteer position filled by a patient (or caregiver of a patient) that receives care at your institution.
* Communicates with patients / families that are new to the CCC
* Gathers feedback from caregivers / patients that receive care at your CCC.
* Shares feedback with VHLA and the CCC Lead Team; meets with the Lead Team at least 1/year to develop improvement plans.

## CCC Sponsoring Physician

* Ensures organization and coordination at the CCC. Designates Point of Contact and VHL Navigator and assembles team of VHL specialists. Shares VHL Surveillance Guidelines within team, ensures team members know each other, and educates team on the importance of patient participation in the MyVHL: Patient Natural History Study.
* Ensures team members notify Patient Navigator when VHL patients are seen in their specialty departments.
* Conducts / facilitates regular communication with entire CCC team in addition to communicating with EMR. Suggestions: manages an internal list-serve for sharing observations about VHL, directs specialists toward VHLA specialty list-serves, encourages specialists to educate their department about VHL, etc.
* Arranges / participates in medical education meetings to teach physicians outside of CCC team about VHL.
* Member of “CCC Lead Team”; meets with VHLA CCC Liaison at least once per year.

## CCC Point of Contact

*Best practice is to designate a genetic counselor or nurse in the sponsoring physician’s department. Consider multiple contacts with a common email address to ensure coverage at all times.*

* Serves as initial contact for patients, families, and physicians. Contact info is listed on VHLA website and given to our hotline volunteers. Uses [VHL Surveillance Guidelines](#_VHLA_Suggested_VHL) to connect patients with relevant CCC specialists. This person should have a basic understanding of VHL (and hopefully [HLRCC](#_Suggested_HLRCC_Surveillance) and BHD), but is not expected to answer any medical questions that require the patient to see a specialist.
* Ensures each CCC Specialists have MyVHL physician information sheets and patient brochures. Frequently encourages patients to participate longitudinally in MyVHL.
* Member of “CCC Lead Team”; meets with VHLA CCC Liaison at least once per year.

## CCC Patient Navigator

* Personally welcomes each VHL CCC patient, introducing them to the concept of comprehensive, coordinated care within the CCC team. Flags patients’ EMR records as VHL and has overview of each VHL patient’s care.
* Works at the patient and institutional level to ensure that each CCC patient is adhering to the [VHL Surveillance Guidelines](#_VHLA_Suggested_VHL) and receiving all recommended tests and treatments.
* When feasible, works with appointment scheduling to expedite and coordinate VHL appointments.
* Has overview of each VHL patient’s care. Follows up with patients that have missed any CCC appointments.
* Monitors the patient’s psychosocial needs using ie: [HADS](#_Suggested_Psychosocial_Questionnair). As needed, works with Specialists to establish relevant care close to the patient’s home.
* Shares genetic testing / surveillance results with patient; ensures patient’s PCP receives report after each CCC visit.
* Reminds / encourages patients after every clinical encounter to participate in MyVHL.
* Determines if patients are facing insurance / financial barriers; utilizes the hospital’s Patient Financial Services Department to assist patients in overcoming these barriers.
* Member of “CCC Lead Team”; meets with VHLA CCC Liaison at least once per year.

## CCC Specialist

* Adheres to [VHL Surveillance Guidelines](#_VHLA_Suggested_VHL). Monitors the patient’s psychosocial needs at every clinical encounter using ie: [HADS](#_Suggested_Psychosocial_Questionnair); as needed, works with Patient Navigator to establish relevant care close to the patient’s home.
* Encourages patients to participate longitudinally in MyVHL after each appointment / procedure.
* Serves as department expert on VHL:
* Receives notification from departmental colleagues each time a VHL patient is seen.
* Ensures VHL Patient Navigator incorporates these patients into the CCC system.
* Educates department on need for coordinated care between multiple specialists to properly manage VHL.
* Understands CCC structure. Knows, works with, and communicates with other team members.
* Becomes a member of VHLA list serve. Posts questions/responses to clinicians at other CCCs.
* Willing to give VHL presentations at physician and/or patient meetings when requested by the Sponsor.

# Application Form: VHL Clinical Care Center

## General Information

**Submission date:** Click here to enter a date.

**Name of person(s) completing this application:** Click here to enter text.

1. The name, address, and main telephone number of the applicant institution:

|  |  |
| --- | --- |
| **\*** Institution | Click here to enter text. |
| Postal Mailing Address | Click here to enter text. |
| Main Telephone Number | Click here to enter text. |

1. Please indicate the patient ages treated at your institution:

Adults only (age 18 years and over)

Pediatric only (under 18 years old)

All ages (*A pediatrician is required to be on the team for all centers seeing children.)*

## CCC Lead Team Information

|  |  |
| --- | --- |
| **\*** CCC Sponsoring Physician | Click here to enter text. |
| Sponsor Specialty | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to academic / clinical bio | Click here to enter text. |

The Sponsoring Physician agrees to the [responsibilities of this position as outlined on page 5](#_CCC_Sponsoring_Physician_1) of this packet.

|  |  |
| --- | --- |
| **\*** CCC Point of Contact | Click here to enter text. |
| Point of Contact Specialty | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |

The CCC Point of Contact agrees to the [responsibilities of this position as outlined on page 5](#_Point_of_Contact) of this packet.

|  |  |
| --- | --- |
| **\*** CCC Patient Navigator | Click here to enter text. |
| Sponsor Specialty | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |

The CCC Patient Navigator agrees to the [responsibilities of this position as outlined on page 5](#_VHL_CCC_Patient) of this packet.

1. What is the professional relationship between the members of the Lead Team—the Sponsoring Physician, Point of Contact and Patient Navigator?   
   Click here to enter text.

## CCC Specialists’ Information

1. For the information of our Clinical Advisory Council, please supply the name of the specialists who have agreed to the [responsibilities of this position as outlined on page 5 of this packet](#_CCC_Specialist). If approved as a CCC, we will add these specialists to our emailing list to receive our newsletter. We will not release their email addresses to patients, but VHLA staff may ask incidental questions via the appropriate VHLA email list serve.

**\*\*Endocrine Surgery**

|  |  |
| --- | --- |
| Physician name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| VHL experience | Click here to enter text. |

**\*Endocrinology**

|  |  |
| --- | --- |
| Physician name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| VHL experience | Click here to enter text. |

**\* Genetics (Genetic Counselor)**

|  |  |
| --- | --- |
| Counselor name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| VHL experience | Click here to enter text. |

**Genetics (Geneticist)**

|  |  |
| --- | --- |
| Physician name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| VHL experience | Click here to enter text. |

**Maternal-Fetal Medicine / High Risk Obstetrics**

|  |  |
| --- | --- |
| Physician name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| VHL experience | Click here to enter text. |

**\*\*Nephrology**

|  |  |
| --- | --- |
| Physician name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| VHL experience | Click here to enter text. |

**Neurology**

|  |  |
| --- | --- |
| Physician name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| VHL experience | Click here to enter text. |

**Neurooncology**

|  |  |
| --- | --- |
| Physician name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| VHL experience | Click here to enter text. |

**\*\*Neurootology**

|  |  |
| --- | --- |
| Physician name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| VHL experience | Click here to enter text. |

**\*Neurosurgery**

|  |  |
| --- | --- |
| Physician name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| VHL experience | Click here to enter text. |

**\*\*Oncology**

|  |  |
| --- | --- |
| Physician name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| VHL experience | Click here to enter text. |

**\*Ophthalmology**

|  |  |
| --- | --- |
| Retinal specialist name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| VHL experience | Click here to enter text. |

**Palliative Care**

|  |  |
| --- | --- |
| Physician name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| VHL experience | Click here to enter text. |

**Pancreatic Gastroenterologist**

|  |  |
| --- | --- |
| Physician name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| VHL experience | Click here to enter text. |

**Pediatrics (\*Required for centers seeing children)**

|  |  |
| --- | --- |
| Physician name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| VHL experience | Click here to enter text. |

**Psychology/Psychiatry**

|  |  |
| --- | --- |
| Physician name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| VHL experience | Click here to enter text. |

**\*\*Radiology**

|  |  |
| --- | --- |
| Physician name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| VHL experience | Click here to enter text. |

**\*Social Worker / Counselor**

|  |  |
| --- | --- |
| Name + position | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| VHL experience | Click here to enter text. |

**Surgical Oncologist**

|  |  |
| --- | --- |
| Physician name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| VHL experience | Click here to enter text. |

**\*Urology**

|  |  |
| --- | --- |
| Physician name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| VHL experience | Click here to enter text. |

#### Additional Specialists with Experience in Caring for BHD / HLRCC Patients

**Dermatology**

|  |  |
| --- | --- |
| Physician name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| BHD / HLRCC experience | Click here to enter text. |

**Gynecology**

|  |  |
| --- | --- |
| Physician name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| HLRCC experience | Click here to enter text. |

**Pulmonology**

|  |  |
| --- | --- |
| Physician name | Click here to enter text. |
| Postal Mailing address | Click here to enter text. |
| Telephone | Click here to enter text. |
| Email | Click here to enter text. |
| Link(s) to bio | Click here to enter text. |
| BHD experience | Click here to enter text. |

## Services Provided

Please indicate whether your medical facility provides the following services on site.

#### Surveillance & Diagnosis

|  |  |
| --- | --- |
| MRI scanner  CT scanner  PET scanner  PET / CT scanner | MIBG  Ultrasound  Audiologic Testing  Genetic Testing |
| Other: Click here to enter text. | |

#### Treatments

|  |  |
| --- | --- |
| Nuclear Medicine  Stereotactic Radiosurgery  Partial Nephrectomy  Open  Laproscopic  Robotic  Resection of Retroperitoneal Paragangliomas | Radio Frequency Ablation  Cryoablation for kidney tumors  Partial Adrenalectomy  Open  Laproscopic  Robotic |
| Other: Click here to enter text. | |

#### Eye Tests & Treatments

|  |  |
| --- | --- |
| Vitreoretinal surgery including vitrectomy  Membrane removal  Endo cyrotherapy | Endo laser treatment  Widefield retinal imagine  Optical coherence tomography  Fluorescein Angiography |
| Other: Click here to enter text. | |

#### Therapy

|  |  |
| --- | --- |
| Physical Therapy  Occupational Therapy  Other: Click here to enter text. | Speech Therapy  Neurological Rehabilitation  Palliative Care |
|  | |

## Short Answer Questions

|  |  |
| --- | --- |
| 1. How many patients with VHL are seen annually? | Click here to enter text. |
| 1. How many patients with HLRCC are seen annually? | Click here to enter text. |
| 1. How many patients with BHD are seen annually? | Click here to enter text. |

1. Why do you feel this institution is a good candidate to be a VHL Clinical Care Center?

Click here to enter text.

1. Beyond electronic medical records, how will your team implement coordinated care?

Click here to enter text.

1. Will you meet regularly as a team? If yes, what will be the frequency?

Yes [ Frequency: Enter Frequency. ]  No

1. How will you update other CCC team members and the patient’s PCP about a patient’s medical status?

Click here to enter text.

1. How will you assist patients in arranging appointments with needed specialists? When feasible, the Patient Navigator should be assisting with this task.

Click here to enter text.

1. How will you assist patients in scheduling their surveillance scans, blood work, etc? What plan does your center have for following up with patients who do not adhere to the surveillance guidelines?

Click here to enter text.

1. Coping with VHL can be an emotional rollercoaster for the patient and loved ones. How will your institution incorporate the emotional issues into the patient’s medical care?

Click here to enter text.

1. How will your site encourage patients to participate in the MyVHL: Patient Natural History Study (vhl.org/MyVHL)?

Click here to enter text.

1. In response to CCCs requesting patient feedback, we have CCC patient liaisons that will meet with the Lead Team (Sponsor, Point of Contact, and Patient Navigator) at least once a year. How else can we support you providing the best VHL care possible? How can we better support you in communicating with prospective patients? All suggestions and feedback are welcome.

Click here to enter text.

1. If there are any existing VHL Clinical Care Centers in your region, how do you plan on working together with them, so as not to offer redundant services and to make sure no patients fall through the cracks? How do you plan on reaching patients that are not already seen by a CCC?

Click here to enter text.

1. Any additional comments in support of the application?

Click here to enter text.

# Clinical Care Center Programmatic Opportunities

## Core Requirements

*These programs are mandatory for VHL Clinical Care Center designation. Please check off each item to confirm.*

**Provide coordinated care for VHL patients**

Hold at least one team meetings of CCC VHL specialists annually

Work with VHLA to identify patient liaison (if one isn’t already in place)

Encourage patients to provide feedback (positive or negative) to VHLA

Implement mechanism for CCC team communication

Distribute Patient Welcome Packet

Assess patient and caregiver emotional health

Participation in/completion of at least 3 of the additional programs/initiatives listed below (dispersed across at least 3 different categories)

## Additional Programmatic Opportunities

*This is a checklist of additional programmatic opportunities. Please check off at least 3 additional opportunities, across at least 3 different categories, that your CCC plans on accomplishing over the next two years. For all opportunities checked, please provide a brief explanation in the space below.*

***Research***

Participate in at least one VHL research initiative or at least one VHL peer reviewed publication in the past 2 years

Submit proposal for VHL specific discussion/talk/presentation at specialty society meeting

Increase and/or maintain MyVHL participation at 50% among CCC patients

***Education***

Host one internal, organizational Grand Rounds program related to VHL, or other internal-facing educational program, formal or otherwise

Send at least one care team member to the Biennial International VHL Medical/Research Symposium

Provide CCC feedback related to areas of improvement for the CCC program to VHLA at least once per year either via form and/or call with VHLA staff/leadership

Submit at least one case to Virtual Tumor Board per year

***Patient Care***

Internal VHL-specific tumor board mechanism and process

Hold at least one patient-facing event annually

Meet with patient liaison once a year

***General***

Create/maintain webpage on hospital website about CCC team, with link to vhl.org

Expand CCC team to include hospital/practice administrative leader

At least 30% care team participation in kick-off meeting (for new and renewed CCCs) plus 100% of core team

***Explanation***

Click here to enter text.

# Suggested Psychosocial Questionnaire

If the center decides to use another questionnaire, please provide a copy and scoring key with the application.

**Revised HADS Questionnaire** – suggested 6 questions

1. **I still enjoy the things I used to enjoy: (depression question)**

|  |  |
| --- | --- |
| 0 | Definitely as much |
| 1 | Not quite so much |
| 2 | Only a little |
| 3 | Hardly at all |

1. **I can sit at ease and feel relaxed: (anxiety question)**

|  |  |
| --- | --- |
| 0 | Definitely |
| 1 | Usually |
| 2 | Not often |
| 3 | Not at all |

1. **I look forward with enjoyment to things: (depression question)**

|  |  |
| --- | --- |
| 0 | As much as I ever did |
| 1 | Rather less than I used to |
| 2 | Definitely less than I used to |
| 3 | Hardly at all |

1. **I get a sort of frightened feeling like ‘butterflies’ in the stomach: (anxiety question)**

|  |  |
| --- | --- |
| 0 | Not at all |
| 1 | Occasionally |
| 2 | Quite often |
| 3 | Very Often |

1. **I can laugh and see the funny side of things: (depression question)**

|  |  |
| --- | --- |
| 0 | As much as I always could |
| 1 | Not quite so much now |
| 2 | Definitely not so much now |
| 3 | Not at all |

1. **I get sudden feelings of panic: (anxiety question)**

|  |  |
| --- | --- |
| 0 | Not at all |
| 1 | Not very often |
| 2 | Quite often |
| 3 | Very Often |

All items scored 0-3 (where 3 is most severe). Alternate items group into Anxiety / depression subscales. Please note that this questionnaire was validated using 12 questions. Totals for each subscale are categorized as follows:

|  |  |  |
| --- | --- | --- |
| 0-3 | Normal |  |
| 4-5 | Mild | Possible clinical disorder |
| 6-7 | Moderate | Probable clinical disorder |
| Above 8 | Severe |

# VHLA Suggested VHL Surveillance Guidelines

If the center uses different VHL surveillance guidelines, please submit a copy of the guidelines with the application.

**Until a cure is found, surveillance is a patient’s strongest defense to prevent severe VHL complications.**[*Revised 4/24/2020]*

Surveillance is the testing of individuals at risk for von Hippel-Lindau disease (VHL) who do not yet have symptoms, or who are known to have VHL but do not yet have symptoms in a particular area. The unaffected organs should still be screened.

Modifications of surveillance schedules may sometimes be done by physicians familiar with individual patients and with their family history. Once a person has a known manifestation of VHL, or develops a symptom, the follow-up plan should be determined with the medical team. More frequent testing may be needed to track the growth of known lesions.

People who have had a DNA test and do not carry the altered VHL gene may be excused from testing.

In order to monitor the most critical areas of the brain and spinal cord in the most efficient and cost-effective manner, CNS MRIs should include the brain, cervical, thoracic, and lumbar spine. Scans should be ordered as no less than a 1.5T MRI with and without contrast, with thin cuts through the posterior fossa, and attention to inner ear/petrous temporal bone to rule out both ELST and hemangioblastomas of the neuraxis.

|  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **Type of Surveillance**  **(Tumors being screened)** | **AGE1** | | | | | | **Pregnancy**10 | |
|  | **Until age 5y** | **Beginning at age 5y** | **Beginning at age 11y** | **Beginning at age 15y** | **Beginning at age 30y** | **Beginning at age 65y** |  |
| **History and Physical Examination**2 | Yearly from age 1y | Yearly | Yearly | Yearly | Yearly | Yearly | Prior to conception10 |
| **Blood Pressure and Pulse**  (Pheochromocytomas/paragangliomas) | Yearly from age 2y | Yearly | Yearly | Yearly | Yearly | Yearly | Prior to conception10 |
| **Dilated Eye Examination**3  (Retinal Hemangioblastomas) | Every 6-12 months, beginning before age 1y | Every 6-12 months | Every 6-12 months | Every 6-12 months | Yearly | Yearly | Prior to conception, then Every 6-12 months10 |
| **Metanephrines**4  (Pheochromocytomas/paragangliomas) |  | Yearly | Yearly | Yearly | Yearly | Stop routine1 | Prior to conception10 |
| **MRI Brain and Spine w/wo Contrast**5,6,7  (CNS Hemangioblastomas) |  |  | Every 2 years | Every 2 years | Every 2 years | Stop routine1 | Prior to conception10 |
| **Audiogram**  (Endolymphatic sac tumors) |  |  | Every 2 years | Every 2 years | Every 2 years | Stop routine1 |  |
| **MRI Abdomen w/wo Contrast**5,6,7  (Renal cell carcinomas, Pheochromocytomas/paragangliomas, Pancreatic neuroendocrine tumors/cysts) |  |  |  | Every 2 years 8 | Every 2 years 8 | Stop routine1 | Prior to conception10 |
| **MRI Internal Auditory Canal**9  (Endolymphatic sac tumors) |  |  |  | Once |  |  |  |

**Notes:**

1 Beginning at age 65, routine laboratory and radiologic screening for patients who have never had specific VHL manifestations may cease. With the exception of routine physical examination and ophthalmologic assessment, this applies to all other routine screening/surveillance tests in asymptomatic patients. However, patients presenting with signs/symptoms should be evaluated with appropriate testing/imaging regardless of age.

2 Age-appropriate history and physical examination to include: Neurologic examination, auditory and vestibuloneural questions and testing, visual symptoms, catecholamine excess symptom assessment (headaches, palpitations, diaphoresis, hyperactivity, anxiety, polyuria, abdominal pain).

3 Dilated, in-person eye examination, including ophthalmoscopy, to occur every 6-12 months based on quality of examination obtained (especially in a child) and perceived adherence to follow-up. Consider examination under anesthesia in young children in whom a detailed eye examination cannot be adequately obtained in the clinic. Consider including ultrawidefield photography and ultrawidefield fluorescein angiography, but these should not replace a dilated eye examination with a specialist with experience in retinal manifestations of VHL.

4 Plasma free metanephrines (preferred, due to its higher sensitivity) or fractionated 24-hour urinary free metanephrines.

5 Use macrocyclic/class II gadolinium-based contrast agents. MRI of the neuroaxis may be obtained at the same time as MRI abdomen, and may be performed under a single long anesthesia event, especially in children. However, both the neuroaxis protocol and the abdominal protocols should be obtained consecutively. It is NOT recommended to evaluate the spine solely using an abdominal protocol MRI, nor is it recommended to evaluate the abdominal organs solely using a neuroaxis protocol. See footnote #6 and #7 for how to combine these protocols.

6 Based on contraindications (metallic implants, renal failure, etc.), the following order of imaging priority applies: MRI (with and without contrast) > MRI (without contrast) > CT (with contrast) > CT (without contrast) > US.(kidneys, adrenals and pancreas only) > Endoscopic US (pancreas only). See also footnote #5 and #7.

7 Timing of contrast administration when imaging multiple organ systems together should be as follows: Obtain non-contrasted images of CNS and abdomen first, then give contrast using a power injector and perform multi-phase contrast-enhanced imaging of the abdomen including pancreas and kidneys during the late arterial phase and delayed venous phases. Then late post-contrast imaging of neuroaxis. See also footnote #5 and #6.

8  If no renal lesions present on initial scan, continue routine surveillance every 2 years. If small tumors (< 3 cm) found, reimage initially with MRI every 3-6 months to determine stability. Once stability has been determined over 3 consecutive scans, consider extending to every 2 years. If renal mass is > 3 cm, consider a referral to a urologist (preferably familiar with the care of VHL).

9 High-resolution (1mm slice thickness) magnetic resonance imaging of the internal auditory canal. This baseline MRI of the internal auditory canal should be obtained after age 15 years (once the temporal bones have matured), and it should be added onto the MRI of the neuroaxis conducted between ages 15-20 years.

10 “Prior” indicates that this surveillance testing should ideally be performed prior to any planned conception, if possible. MRIs performed during pregnancy should be   
 without contrast.

# Suggested HLRCC Surveillance Guidelines

*Approved by HLRCC Clinical Advisory Council*

If the center uses different HLRCC surveillance guidelines, please submit a copy of the guidelines with the application.

The suggestions in this book come from the most experienced research projects, which still are only 10 years old, and therefore do not yet have long-term follow-up experience. Some physicians are of the opinion that there is insufficient scientific data yet to define firm recommendations.

Recommendations presented in this handbook should be regarded as tentative, and will most likely change over time as more data becomes available. We suggest these guidelines as a starting point, and ask for feedback to help shape better guidelines in the future.

The one common guideline we do recommend is that you take charge. Be informed, discuss openly with your medical professionals and consider the pros and cons. Please e-mail us at info@hlrccinfo.org if you have unanswered questions and we will always try to help you.

HLRCC is a relatively young condition in that there are few people who have been followed medically from childhood through adulthood with today’s surveillance technologies. We have lots of questions, but few firm answers at this time. However everyone with HLRCC can assist us in learning what we need to know to protect ourselves and our children from the worst effects of HLRCC.

The purpose of surveillance guidelines is to help patients and their local physicians watch out for foreshadowing of problems, before they get to a critical stage. By intervening at earlier stages, hopefully these problems never become threatening to life or quality of life. Note that surveillance means that you do not yet have any issues in the area being screened. Once you have a diagnosed issue, then you will need to follow the guidance of your health care team in deciding what course of action to take. Feel free to seek a second opinion from a doctor more familiar with HLRCC, even in another country. Scans can easily be sent through the mail, and the experts are quite willing to provide their opinion. Note: e-mailing scans is not satisfactory. There are many file formats used by the various vendors, so the receiving doctor will likely not be able to read the file. On the CD they include the viewer software to read the file, so the receiving doctor can open the file and read it quite nicely. Sending a universal format (like a .pdf) reduces the quality of the image, making it hard to determine what is a cyst and what is a tumor—a critical distinction for us.

* MRI is recommended in order to minimize exposure to radiation.
* CT should be reserved for times when they are needed to answer some specific diagnostic question or in planning surgery.
* Ultrasound of the kidney is not recommended, as ultrasound is very much dependent upon the quality of the machine and the skill of the operator. Ultrasound is better than nothing, but is unlikely to find tumors less than 1 cm, leaving a wide opportunity for risk in HLRCC.

The surveillance guidelines shown in this section are the best advice we were able to assemble at this time from the major research teams studying this condition, and from patient experience. We are hopeful that using these guidelines as a starting point, and with feedback from patients and physicians worldwide, we can evolve these guidelines over the next several years to make them increasingly cost-effective.

##### For Children at Risk Under Age 8

Children are “at risk” if they are not genetically tested or if they are in the 3% of families which have clear evidence of HLRCC symptoms, but no DNA alteration can be found.

#### Annually from age 1

* Full body skin check by the pediatrician, noting any skin bumps. If present, refer to a dermatologist familiar with HLRCC

#### Annually beginning at age 8

* Full body skin check by the pediatrician noting any skin bumps. If present, refer to a dermatologist familiar with HLRCC
* MRI of the abdomen with thin cuts to check for any cysts or tumors of the kidneys. If present refer kidney issues to an urologist familiar with HLRCC.

##### Age 8+

* Full-body skin check by a dermatologist to note the location, number, and stage of skin bumps
* MRI of the abdomen with thin cuts looking for kidney tumors, noting number and size of any cysts or tumors seen
* Adults may be offered a CT scan as well as an MRI for their first appointment in order to have a baseline scan for comparison with any later scans
* For women starting at the age of 21, annual examinations by a gynecologist to enable surveillance for fibroids. You should inform the gynecologist that you have the HLRCC condition, of any family history of fibroids and stress the importance of looking for even small fibroids.

# Suggested Referral Criteria to VHL Clinical Care Center

*Developed and Used by Othon Iliopoulos, MD****\*,*** *Massachusetts General Hospital, Boston MA*

|  |
| --- |
| **1.  Any blood relative of an individual diagnosed with VHL disease.** |
| **2.  Any individual with TWO VHL-associated lesions**  **\*VHL associated lesions:** hemangioblastoma (HB), clear cell renal carcinoma (RCC), pheochromocytoma (PHE), endolymphatic sac tumor (ELST), epididymal or adnexal papillary cystadenoma, pancreatic serous cystadenomas, pancreatic neuroendocrine tumors. |
| **3.  Any individual with ONE or more of the following:**   * CNS hemangioblastoma * Pheochromocytoma or paraganglioma * Endolymphatic sac tumor (ELST) * Epidiydmal papillary cystadenoma |
| **4.  Any individuals with**  Clear cell renal carcinoma (RCC) diagnosed at a < 40 year old patient Bilateral and/or multiple clear cell RCC  >1 pancreatic serous cystadenoma >1 pancreatic neuroendocrine tumor Multiple pancreatic cysts + any VHL-associated lesion |

*These are criteria used to REFER patients. These are NOT criteria for clinical diagnosis of VHL.*

\* Based on Melmon KL, Rosen SW, Lindau’s Disease: Review of the literature and study of a large kindred. Am J Med 1964; 36:535-617.