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CRITERIA FOR AN INTERNATIONAL CURE HHT CENTER OF CARE

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DEFINITION OF AN ICOC

A Hereditary Hemorrhagic Telangiectasia (HHT) International Center of Care (ICOC) is a medical team outside of North America, recognized by Cure HHT as possessing the best available personnel, interest, expertise, commitment, and resources in their country or geographic region to evaluate, treat, and educate families with HHT.

GUIDING PRINCIPLES

These criteria were developed by the Cure HHT Centers of Excellence committee with the following guiding principles in mind:

- HHT is underdiagnosed around the globe
- There are few physicians with interest in and/or experience with HHT in many regions of the world, leaving many people with HHT undiagnosed or in need of education regarding their disease and options to treat the manifestations of HHT
- Patients are in need of physicians that have an interest in the diagnosis and treatment of HHT. Recognizing ICOCs will facilitate people with HHT finding a center closest to them that is able to provide care tailored to HHT.
- Recognizing Centers of Care will allow more physicians to develop higher degrees of expertise in the diagnosis and treatment of HHT patients
- Developing ICOCs is important to help connect HHT experts in other countries to increase knowledge and expertise which will further improve patient care in many parts of the world
- Cure HHT does not have the resources or obligation to fully vet physicians, hospitals and/or institutions in other countries during the process of designating an ICOC
- Unlike North American COEs where there are standards of care that are similar for each region, there exists a very wide variability of knowledge and resources in other countries. With this in mind, designating ICOCs in countries outside of North America must take into account other potential options for management of persons with HHT in that region. Therefore, standards for an ICOC may be more rigorous in countries where there are options for “advanced” HHT care (i.e. pulmonary arteriovenous malformations (PAVM) embolization) compared to other countries where the mere presence of a physician who has basic knowledge and interest in HHT

may be the only resource available to patients in that country or region. Given this important difference, criteria for each ICOC must be compared not to ICOCs in other countries/areas, but between other potential options in each country/region.

- The ultimate goal for the establishment of an ICOC is to further develop and provide expert level of care among the subspecialties required to best help diagnose and treat people with HHT.

GENERAL ICOC EXPECTATIONS

ICOC are encouraged to:

- Educate patients, families, and health care providers about HHT
- Interact with the non-ICOC health care providers of persons with HHT
- Engage in HHT research
- Distribute Cure HHT education materials
- Participate in Cure HHT activities such as webinars, conferences, Listserv, etc.

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ICOC CRITERIA

The required criteria to initially obtain and maintain ICOC certification are detailed below in 4 sections:

- A. Personnel
- B. Facilities and Resources
- C. Education, Research, and Advocacy/Administration
- D. Other ICOC expectations

A. Personnel

ICOC should have a designated person with both an interest and knowledge of HHT in order to provide a level of care that may not be readily available elsewhere in the community. At most successful ICOC, specialty services are best served by one clinician as opposed to a group of clinicians (e.g. one ENT physician as opposed to several). This is because the experience of managing a rare disease can be focused on one clinician and allow for development of greater expertise in contrast to diffusing that experience over a group. Spreading the experience across a group is most appropriate when a senior clinician is training his or her replacement or with an established ICOC/COE that has a large volume of patients. Of course, it is appropriate to have back up clinicians who can fill in when the main specialist is out of town or in case of other emergencies. If available, it may also be advantageous to have a second physician who could be mentored by a more senior person as well.

Core Staff

Ideally an ICOC should have the following core staff available to diagnose and treat patients with HHT. This core staff would be expected to be present and available at larger ICOCs where such staff with an interest and/or expertise in HHT is otherwise available within the country or region. For smaller ICOCs where such resources are not practically available, only a medical director and

coordinator may be available to see HHT patients. However, it should be a goal to increase the number of core staff available over time as is practically possible given the limitations that may be present in the region and providing genetic testing if possible.

The following personnel must be designated and available at an ICOC.

1. Medical Director. The Medical Director must have specialized knowledge of the main organ manifestations of HHT and experience (or in some cases an interest in obtaining experience) in their management. An important part of this role is having the time and ability to coordinate the multidisciplinary diagnosis and treatment of patients referred to the ICOC. If a multidisciplinary diagnosis and treatment team is not available, it should ideally be a goal for such a center to be developed. The Medical Director is expected to see at least 90% of patients new to the ICOC and serve as the “HHT primary provider” for issues related to HHT (not to be confused with the patient’s primary care physician (PCP) who manages all things not HHT). If at some point the volume of HHT patients exceeds what one person could reasonably manage, one or more colleagues may need to assist the Medical Director in his/her role as the HHT primary provider. Serving as the HHT primary provider includes spending time devoted to: seeing patients in clinic, screening for visceral manifestations, following patients on other services while they are receiving inpatient care at the ICOC, and dealing with complex issues leading to referral and follow-up.

For more advanced ICOCs, the Medical Director position for a new ICOC cannot be evenly shared between co-directors, though an Associate Director is encouraged. For example, a junior clinician with limited experience in HHT might be appointed as Medical Director as long as he or she has a senior clinician with extensive HHT expertise acting as mentor in the role of Associate Director. As another example the Medical Director might be adult focused while the Associate Director is pediatric focused (or vice-versa).

Criteria include:

- a. A degree of MD (or equivalent)
 - b. Available time commitment to see the majority of all HHT patients presenting to the ICOC
 - c. Engagement with the HHT community through conferences, webinars, Listservs, etc. depending on individual circumstances
 - d. Inclusion in the Cure HHT physician directory
2. Coordinator. A coordinator should be included as an ICOC staff member. At many centers, this position is filled by a nurse. For small ICOCs, this role may be assumed by the HHT physician if needed. The Coordinator is typically responsible for the initial patient contact - determining what tests and consults are necessary for the initial visit to the ICOC and coordinating all aspects of that visit. Accordingly, the Coordinator must have

sufficient knowledge of HHT to make these decisions and must be able to respond to patient inquiries in a timely fashion.

3. Otolaryngologist (ENT). Ideally an ICOC should have an ENT physician with interest and experience in the treatment of recurrent epistaxis in HHT patients. Optimal management of epistaxis in HHT requires a collaborative effort that includes surgical and medical management. It is understood that in some regions of the world this may not be possible and available surgical treatments may be limited or even non-existent at the time of ICOC formation. It is hoped that by forming an ICOC in these places a sufficient patient volume may develop which would allow for such services to be developed. In such places, the Medical Director may initially provide all epistaxis treatment with local measures and/or with systemic medications.

The ultimate goal for ENT service capability would include the following:

- a. Experience with advanced cautery techniques (including bipolar cautery, laser cautery, and coblation)
 - b. Experience with and/or referral pathways for advanced procedures to treat epistaxis such as sclerotherapy, Young's nasal closure, or nasal septal dermoplasty
4. Interventional Radiologist (IR). Pulmonary AVMs are a major source of morbidity and mortality. Therefore, it should be the goal of each ICOC to be able to properly screen, diagnose and treat PAVMs. The ability to treat (or refer for treatment) PAVMs is a requirement for an ICOC where PAVM treatment is readily available in the region. Ideally this physician would have experience and expertise in the treatment of pulmonary AVM. The IR should also have an understanding of the complexities of treating PAVM such as the treatment of pregnant patients and re-perfused PAVM.
 5. Hematologist. As greater than 50% of HHT patients are anemic, the availability of a hematologist with the understanding of the unique needs of HHT patients is very helpful. In the absence of such a physician, the Medical Director or other physician who is knowledgeable about iron replacement and the use of medications to alter bleeding in HHT patients can also be utilized.

Additional Staff

In addition to the core staff above, an ICOC should ideally have ready access to the following personnel if available in their region. Referral outside the specific ICOC should be made if possible for services not available at the ICOC institution. It is recognized that many ICOCs may have few, if any, of these services that are readily available. ICOCs in regions of the world where these services are obtainable would be expected to have these.

1. Neurovascular team. Typically, a team of Interventional Neuroradiologist and Neurosurgeon with experience and expertise in the diagnosis and treatment of brain and spinal cord AVM and other cerebrovascular lesions.

2. Genetic counselor or Geneticist. This person should have expertise to provide HHT-specific genetic counseling to patients and families.
3. Pulmonologist. This person should have experience in the management of pulmonary AVM and their complications, including hemoptysis.
4. Cardiologist. This person should have experience in the management of cardiovascular HHT complications including pulmonary hypertension, high output heart failure, and atrial fibrillation.
5. Dermatologist and/or Oral Surgeon/Dentist. This person/s should have experience in the management of non-nasal mucocutaneous telangiectasias.

B. Facilities and Resources

An ICOC should have the facilities and resources required to perform all the procedures necessary to evaluate and treat the manifestations of HHT whenever possible. In regions of the world where these services are generally available, it would be expected that the ICOC would be able to provide these services or have relationships with physicians and/or institutions that could provide them.

Criteria include facilities to perform the following:

1. MRI imaging of the brain
2. Agitated saline contrast echocardiography
3. CT imaging
4. Interventional radiology services for pulmonary angiography and percutaneous embolization therapy
5. Advanced cautery equipment such as KTP laser, YAG laser, bipolar electrocautery, and/or coblation
6. Iron infusion capabilities
7. Access to obtain genetic testing for the main HHT-causing mutations
8. Adequate record keeping system
9. Access to gamma knife for treatment of cerebral vascular malformations is encouraged but not mandatory
10. Access to capsule endoscopy and extended upper GI endoscopy (e.g. double balloon enteroscopy) is encouraged but not mandatory
11. Right heart catheterization

C. Education, Research, and Advocacy/Administration

Patient care and understanding of HHT are optimized when ICOC/COE are collaborating with each other and with Cure HHT to provide education about HHT, conduct research, and participate in a wide spectrum of advocacy and administrative activities. Accordingly:

- Centers applying for initial certification are encouraged to progressively participate in the following activities, but these are not required criteria for initial certification.

1. Education

Educational activities are an important component of a ICOC and are necessary to maintain an up-to-date understanding of the management of HHT and to inform others about HHT.

Excellence in Education should be demonstrated by one or more staff fulfilling criteria (when possible) which may include:

- a. Participation in the Cure HHT global Listserv
- b. Attending Cure HHT webinars
- c. Attendance at HHT clinical and/or research meetings either in person or virtually when available
- d. Participation in HHT related research

D. Other ICOC Expectations

1. Patient volume

Patient volume expectations will vary based upon the current expertise of the ICOC staff, the number of HHT patients currently thought to be diagnosed and/or treated in the region and other factors. It is expected that the volume will grow as the ICOC matures.

2. Timeliness of communications

Patients and families should expect timely correspondence regarding appointments, results of testing and need for specific treatments.

It is expected that the ICOC will respond timely to Cure HHT communication efforts.

3. Staff changes

All ICOC must notify Cure HHT of changes in Core Staff within 30 days.

4. Annual Status Reports

Each ICOC should submit an annual status report that will include a limited data set such as personnel changes, number of patients seen, number of PAVM embolizations, etc.

CENTER IDENTIFICATION PROCESS

The above criteria cannot foresee every possible circumstance that may occur at prospective or established ICOC. Therefore, the Cure HHT ICOC Committee reserves the right to be judiciously flexible in the interpretation of these criteria when reviewing applications for initial ICOC certification and established ICOC for recertification.

Initial Identification

- In general, Cure HHT discourages more than one ICOC in the same geographic area. Given that development of expertise in the management of HHT is almost certainly correlated with patient volume, and since HHT is a rare disease, 2 ICOC in the same geographic area would likely not maintain sufficient volume.

Process of Initial Identification

Identification of a potential ICOC could be based upon contact by the potential physician with an interest in HHT or via a referral from an HHT contact from within the global HHT community

Applicants interested in becoming a ICOC should thoroughly review the above ICOC criteria and then contact Cure HHT to express an interest in being designated as a center.

A potential ICOC should identify, or ask for recommendations, a COE that can serve as a mentor institution during the development of their program. It would be anticipated that this mentor center would keep in contact during the early phases of the both the application process and the start of the center after designation. Upon receipt of the expression of interest, Cure HHT will refer the potential center to the ICOC committee for further review and action which will include contact with the proposed Medical Director and other staff members when available. Once the ICOC committee has obtained information regarding the proposed center, the staff and reviewed this data with regard to other available resources in the region, and then the committee will present their findings to the full center committee for further discussion and further action as needed.

Outcome of Initial Certification Evaluation

After full center committee review a written summary of the center evaluation will be provided to the Applicant. Possible outcomes of the evaluation are outlined below.

1. Certification granted
2. Certification is on hold and details needed to move forward will be discussed

Recertification

The overarching goal of the recertification process is to ensure that ICOC are providing comprehensive, state-of-the-art care to HHT patients. This process will provide a mechanism to ensure that all Centers continue to meet the needs of the region served by the ICOC. It will also provide an opportunity for ICOC to help each other provide HHT care that meets our common, agreed-upon standards.

Recertification of established ICOC will occur at regular intervals (~5 years).

Recertification will take place under the guidance of the ICOC committee which will be looking to see that the region is benefitting from the presence of the center and, ideally, the resources and abilities of centers in developing areas are growing over time.