

# The Barrow Hypothalamic Hamartoma Program

---

Maggie Varland, RN, BSB

---

*The Hypothalamic Hamartoma Program falls under the Skull Base Center at Barrow Neurological Institute. The multidisciplinary team is designed to provide comprehensive evaluation, diagnostic testing, and treatment for patients with hypothalamic hamartomas. The team conducts patient care conferences, supports collaboration between clinical and research specialists, and facilitates educational programs to advance the diagnosis and treatment of this complex disease process. This subspecialty program provides patients the latest technological and medical advances and unites clinicians from numerous disciplines to treat patients seeking curative or palliative care for their condition.*

**Key Words:** hamartoma, hypothalamus, skull base

The Barrow Neurological Institute of St. Joseph's Hospital and Medical Center provides the latest technology and medical advances to patients seeking curative or palliative care for their condition. The vision of the institution is to become the premiere neurological treatment center in the country for the comprehensive management of patients with diseases of the brain, spinal cord, and peripheral nervous system. Barrow seeks to remain a leader in neurological research, community education, and quality patient care by creating a comprehensive medical center that encompasses a variety of subspecialties.

The subspecialty programs unite clinicians from numerous disciplines to treat patients with conditions that require complex diagnostic testing, treatment planning, and evaluation. The Hypothalamic Hamartoma Program represents a unique new center at Barrow. These lesions produce symptoms that affect neurological function in the form of epilepsy. They also affect the endocrine system and neuropsychological development. Unless medical professionals assess such patients systematically rather than by seizure evaluation alone, hypothalamic hamartomas can be misdiagnosed or go undiagnosed completely.

Nationally, this disease appears to have a prevalence of one to two cases per million population. This number, however, is quite likely an underestimate of the true prevalence of the condition because these lesions can be overlooked on brain imaging studies. Furthermore, the number of adults living with this disease who are suffering

---

*BNI Administration, Barrow Neurological Institute, St. Joseph's Hospital and Medical Center, Phoenix, Arizona*

from seizures, rage behavior, and growth abnormalities is unknown. This article describes the Hypothalamic Hamartoma Program at Barrow, which is the only treatment center in the United States providing comprehensive service to individuals with hypothalamic hamartomas.

## The Multidisciplinary Team

The Hypothalamic Hamartoma Program is composed of a multidisciplinary team designed to provide comprehensive evaluation, diagnostic testing, and treatment for patients with hypothalamic hamartomas. The team includes primary and support members. The need for each specialty depends on the age and specific medical issues affecting each patient.

Each team member's role is defined, but their responsibilities vary as the needs of the patient evolve throughout the course of therapy. The primary team members take an active role in the planning, implementing, and evaluation of the treatment plan for each patient. These team members include a neurosurgeon, neurologist, endocrinologist, radiation oncologist, neuroradiologist, and neuropsychologist. Other members essential to the success of the team include a nurse practitioner and a nurse coordinator who provide educational and psychosocial support to patients and family members. Support team members include other individuals who may interact with the patients at various stages of care: ophthalmologists, anesthesiologists, geneticists, and rehabilitation specialists.

### Role of Nurse Coordinator

The program coordinator functions as the centralized contact to provide information and direction to the team. This information is shared through a formal communication system established to facilitate collaboration. The documentation may assume the form of progress notes, a dictated letter, or a formal team report. Collaboration through the team conferences and centralized

communication path significantly improves the efficiency, quality, and continuity of care.

The program coordinator serves as the facilitator for the team and has the responsibility of assessing many of the team's functions, such as interactions and proceedings during the course of a conference as well as the overall accomplishments of the program. The focus of the program coordinator has broadened to include evaluation of the team's goals such as timeliness of completion, conformance to standards set by the team, resources consumed, and patient satisfaction. Other responsibilities include acknowledging, encouraging, and supporting each team member while remaining available to them.

As the patient liaison, the program coordinator performs a variety of functions. She coordinates surgical schedules and team consultations and orchestrates the review of patients' medical records. Considerable time is devoted to educating patients and families about their individualized treatment plan and about what to expect at each stage of their care. Family participation is strongly supported, and family members are encouraged to ask questions and to voice their concerns. The coordinator performs extensive follow up with patients to ensure the continuity of their care and successful outcomes. The coordinator also provides information to families about local community resources available to meet the patient's medical and family's personal needs.

### Role of Neurology

Antiepileptic drugs (AEDs) may control the seizures of some patients with a hypothalamic hamartoma. More typically, however, AEDs offer limited or poor control with respect to seizure management. As a result, many children with this condition suffer multiple gelastic seizures daily.

The Division of Neurology at Barrow has one adult and two pediatric epileptologists who are trained in the latest epilepsy treatments, including surgery, vagal nerve stimulation, and

ketogenic diet. Before surgery or Gamma knife treatment, all patients are evaluated by one of the epileptologists, who obtains a detailed history of their seizures and current AEDs. Blood tests, AED levels, and other investigations may be performed. If the patient has a vagal nerve stimulator, it is turned off by the neurologist before the patient undergoes preoperative magnetic resonance imaging and surgery. In all cases, an epileptologist reviews the patients' records before the patient's initial consultation. At times specific recommendations are made (and discussed with the patient's primary neurologists and caregivers) before the initial consultation. For example, the use of divalproex sodium may be discontinued before surgery (to avoid the slightly increased risk of bleeding associated with its use), or a trial of another AED may be recommended if only one or two AEDs have been previously tried. Postoperatively, a neurologist follows the patient and monitors the use of AEDs throughout the patient's hospitalization.

### Role of Neurosurgery

Neurosurgical intervention is indicated only after radiographic and physical examinations confirm the diagnosis of hypothalamic hamartoma. When patients with hamartomas fail to respond to medical treatment and their seizure condition is sufficiently severe, the lesion can be treated surgically.

Preoperatively, the surgeon's role includes assessment of the patient's clinical status and of the size and anatomy of the lesion. Only the surgeon can define the specific risks involved in surgical intervention in an individual case for patients and their families. It may be impossible to resect small hamartomas associated with both a seizure disorder and precocious puberty from a standard transcallosal approach, and an alternative approach such as the orbitozygomatic approach may be necessary. However, complete or near complete transcallosal resection of hamartomas can usually be achieved relatively safely. Details regarding neuro-

surgical management and the anticipated outcomes are discussed elsewhere in this issue.

### **Role of Stereotactic Radiosurgery**

Gamma knife therapy is available at the Barrow if this option proves to be the best treatment for an individual patient. When Gamma knife therapy is selected, a treatment plan is developed, delineating the hypothalamic hamartoma as the target. A dose of radiation is delivered to the hamartoma while avoiding excessive radiation to the optic chiasm and adjacent vital structures. During Gamma knife treatment, the patient is exposed to 201 very thin beams of radiation that only intersect in the target. During this procedure, adults are usually sedated while children are placed under general anesthesia. It is important to recall that the therapeutic effect of Gamma knife is delayed for as long as 6 months after treatment.

### **Role of Endocrinology**

Some patients with hypothalamic hamartomas that cause epilepsy also may have precocious puberty or other signs of a comorbid endocrinological disturbance. Surgery involving the hypothalamus can profoundly change the hormonal status of a patient by causing problems, usually temporary, with the regulation of fluid balance, hunger, growth, and response to stress. Ideally, an endocrinological evaluation should be completed in the patient's home community at the time of initial diagnosis. The role of the endocrinologist on the Barrow multidisciplinary team is to evaluate the patient's hormonal status before surgery, to assess the effects of surgery on the patient's hormonal status in the postoperative period, and to plan for follow up for late assessment of the patient's hormonal status after full recovery.

### **Role of Neuropsychology**

Individuals with a hypothalamic hamartoma can have a wide variety of cognitive and behavioral difficulties, and each patient's level of cognitive

functioning must be understood before surgical intervention. Consequently, different types of neuropsychological tests might be administered, depending on the individual needs of a patient. These tests provide an objective analysis of a patient's neuropsychological status before and after surgery. The patient's family completes questionnaires concerning the patient's level of competence in daily life, personality, and behavioral characteristics. Adult patients fill out similar questionnaires.

Hypothalamic hamartomas may affect cognitive and personality function in different ways, depending on a patient's age and how long the tumor has influenced normal neurological development. Typically, patients exhibit some limitations in intelligence, problem-solving ability, and emotional and motivational development. By understanding the nature of the individual's neuropsychological status, family, teachers, and patients themselves can better understand why adjustment problems exist and how they may best be managed. The patients and families are often perplexed about the cognitive and personality changes associated with this neurological condition. Discussion about the nature of the disturbance often helps reduce confusion about why a given behavior or cognitive problem is present. This information can lead to practical suggestions for management and education.

### **Team Synergy**

The individuals on the hypothalamic hamartoma team seek challenges, want to make a contribution, are assertive, and enjoy collegiality and working with the latest technology. They value autonomy and take pride in their profession. Mutual respect, flexibility, setting goals, sharing both success and tribulations, and recognition of everyone's contribution to the care of each patient create a positive environment where creative, quality patient care can flourish.

The team's goals, and priorities, which are clear and specific, consis-

tently focus on the patient. Although a successful outcome is the team's ultimate goal, providing emotional and spiritual support to patient and family is critical. The values the group embraces include faith, compassion, personal responsibility, respect, listening, understanding, and trust.

The team solves problems systematically through appropriate discussion. They define and address issues of contention rather than criticize individuals. The team members appreciate and respect the different perspective that each member offers. When individuals with highly respected levels of expertise are involved in creating a care plan, the solutions they develop are more widely accepted by the patients as well as by the medical community than if created by one person alone. This dynamic is critical to the success of the program.

### **Program Process**

The team conducts monthly patient-care conferences, supports collaboration between clinical and research specialists, and facilitates educational programs to advance the diagnosis and treatment of hypothalamic hamartomas. The team reviews all pertinent medical records to fashion a treatment plan to meet the specific needs of each patient based on each member's expertise and experience and on the consensus of the group.

When a patient is referred to the team, whether by self-referral or by their physician, the program coordinator collects pertinent medical records and copies of all radiological studies. This information is disseminated to the team members for their evaluation. After reviewing the available records, a neurologist or neurosurgeon presents each patient to the team to ensure that the case is discussed in an effective manner.

Each patient must be assessed to devise an individualized treatment plan that can achieve the goals mutually agreed upon by the patient, family, and medical team. The plan of care is out-

lined and coordinated with the patient and each team member who will be involved in the care of that individual. The role of each specialist is detailed and clearly defined.

After each patient review conference, team reports are processed by the program coordinator and sent to the other team members while the patients are contacted to discuss the team's recommendations. The patients are provided with a directory, which includes the contact information for all team members, should patients want further information about a particular issue. The team often serves as a reference for patients and other medical professionals seeking information about this rare disease.

Once the relationship between the team and patient is established, the program coordinator discusses the family's critical role in the patient's treatment. The family's efforts and contributions are acknowledged and recognized as equally valuable as the efforts of the health care team. Preoperative teaching for the patients and their families includes discussion of the disease process;

the events before, during, and after surgery; discharge planning; prevention of complications; resource availability; and individual patient needs. Hospital resources for family support, such as social workers, chaplain services, an admissions representative, and community resources, are identified for the family and access to these resources is facilitated.

The program coordinator continues to work closely with each patient and their family during the hospital stay. Additional responsibilities include ensuring proper communication with the patient's local physicians at the time of discharge, overseeing the collection of follow-up data, and managing the patient database.

### Support Groups

In addition to their physicians, patients with a rare disease may seek assistance for the direction of their care from a regional or national support group. The Hypothalamic Hamartoma Support Group (HHUGS) offers individuals afflicted with this disease support and information and can serve as

an invaluable resource (<http://www.geocities.com/hhugs2001/>).

### Conclusion

To date, 30 patients with hypothalamic hamartomas have been treated at Barrow. One patient received Gamma knife therapy while the remaining 29 underwent surgical intervention. So far their outcomes have been promising. The hypothalamic hamartoma team is currently exploring treatment options for patients from all over the United States, and internationally.

For children and adults with hypothalamic hamartomas to have productive futures, health care providers throughout the world must not only be aware of this disease but must recognize the symptoms early in the disease process. Barrow is committed to educating the community and other health care professionals. Web site, publications, national presentations, brochures, and direct correspondence with patients and their local physicians are in progress.