

MRI-Guided Laser Ablation Surgery of Hypothalamic Hamartomas

HOW DOES THE TEAM DECIDE IF A PATIENT IS A CANDIDATE FOR MRI-GUIDED LASER ABLATION?

A careful review of each patient's medical records is the first step, including MR imaging of the brain and any applicable neurology or neurosurgery records. Patients with Hypothalamic Hamartomas (HH) typically have gelastic seizures, which are characterized by emotionless laughing, although variations including abnormal movements or staring spells are also common. Every patient's case is handled individually, and it may be necessary for a patient to come to Texas Children's Hospital for further testing to determine if they are a candidate for MRI-guided laser ablation surgery.

WHAT HAPPENS DURING MRI-GUIDED LASER ABLATION SURGERY?

After being placed under general anesthesia, a head frame, or a set of markers, is fitted to the patient's skull. A CT scan is completed to orient the brain to the frame in 3 dimensions. With the help of computer software, a safe pathway that goes through the brain to the HH is calculated for the laser. The neurosurgeon then makes a small incision and drills a small hole through the skull (3.2 mm wide). The laser applicator, a small tube about the width of a strand of spaghetti, is inserted and guided through the brain into the HH. Once the laser applicator is inserted into the brain, the head frame is removed, and the patient is transported to the MRI scanner. After confirming proper placement of the laser applicator and setting safety markers, the surgeon performs a small test firing using the laser. Once accuracy is confirmed, the laser is heated to destroy the HH. Once completed, the applicator is taken out, and the scalp is closed with a single stitch. The patient is then brought to recovery.

HOW LONG DOES MRI-GUIDED LASER ABLATION SURGERY TAKE?

Patients typically spend 6-8 hours under general anesthesia. The majority of this time is surgical planning and MRI scanning after the laser applicator is placed. The actual ablation itself, when the laser is turned on and HH tissue is destroyed, only takes minutes. Throughout a patient's surgery, the family will stay in the surgical waiting room and will be updated regularly by the operating room patient care liaisons.

WHAT TESTING IS NEEDED BEFORE MRI-GUIDED LASER ABLATION SURGERY?

An EEG will be performed to capture seizure activity. Some patients may need an overnight EEG, which requires an inpatient stay in the Epilepsy Monitoring Unit (EMU). Special imaging of the brain is completed including an MRI scan, PET scan, and resting-state functional MRI scan. As imaging protocols can vary widely among institutions, these tests must be obtained at Texas Children's. The PET scan is a nuclear medicine study which demonstrates the metabolism of the HH. The resting-state functional MRI identifies network connectivity within the brain and is used to identify normal and abnormal connections to the HH. In addition, all patients are required to undergo a thorough evaluation with a neuropsychologist prior to surgery (in non-verbal children or children under 3, a developmental evaluation is ordered). Families that are traveling from out of state or outside of the U.S. should have neuropsychological testing completed locally prior to arriving at Texas Children's for surgery. Appointments with an epileptologist (a neurologist that specializes in the treatment of epilepsy) and a neurosurgeon will be arranged prior to surgery. In addition, a thorough endocrine evaluation should take place prior to surgery, as some patients with HH may have endocrine abnormalities.

HOW DOES THE TEAM DECIDE WHAT AREA OF THE HH TO ABLATE?

The goal of MRI-guided laser ablation surgery is to destroy as much of the HH as possible and disconnect it from normal brain tissue. Prioritization is guided by results of the PET scan and resting-state functional MRI, both of which help to look at the most active areas of the HH.

HOW DOES THE TEAM DECIDE WHERE TO PLACE THE LASER APPLICATOR TO ABLATE THE HH?

A careful review of each patient's pre-operative imaging is completed. Using special high-resolution software that reconstructs images of the patient's brain in 3-D, the surgeons plan a path to the HH that avoids blood vessels and damaging or passing through any vital structures while maximizing the area of the HH to be destroyed and/or disconnected.

WHAT HAPPENS TO THE HH AFTER MRI-GUIDED LASER ABLATION SURGERY?

The HH tissue is consumed by the immune system. It sometimes leaves a fluid-filled space behind or an area of brain scar, or “gliosis”, neither of which are likely to cause seizures to return.

CAN THE HH RETURN AFTER MRI-GUIDED LASER ABLATION SURGERY?

The HH is a malformation in normal brain growth that occurs during fetal development. The HH grows in proportion to a person’s normal brain growth, and does not grow after 10 years of age. It will not spread to other areas of the brain or body. HH tissue does not grow back after MRI-guided laser ablation.

WHAT ARE THE SEIZURE-FREE RATES OF PATIENTS THAT HAVE HAD MRI-GUIDED LASER ABLATION SURGERY AT TEXAS CHILDREN’S HOSPITAL?

Currently, 93% of patients are seizure-free at 1 year after MRI-guided laser ablation surgery at Texas Children’s Hospital.

CAN SEIZURES RETURN AFTER MRI-GUIDED LASER ABLATION SURGERY?

Seizures that occur within several weeks of MRI-guided laser ablation are not alarming because there is a network within the brain’s neurons associated with the HH which can independently fire. This does not mean that the surgery has failed. Seizures that persist beyond a few weeks are also not alarming as long as they are different from a patient’s seizures before surgery (shorter, less frequent, causing less after-seizure effects, etc.). In patients with multiple daily seizures, there is a process that typically occurs after MRI-guided laser ablation surgery that is called a “running-down” phenomenon. During the run-down, the networks within the brain that used to spread seizures sometimes activate by themselves, causing seizures to occur. These networks take up to 9 months to quiet. All epilepsy surgery centers assess their outcomes of epilepsy surgery at 1 year after surgery due to this phenomenon. Many children are immediately seizure-free upon the completion of MRI-guided laser ablation and do not experience a seizure run-down effect.

WHY MIGHT SEIZURES RETURN AFTER MRI-GUIDED LASER ABLATION SURGERY?

After MRI-guided laser ablation surgery, seizures may return for a variety of reasons. Sometimes the entire HH may not be able to be destroyed with the laser due its size or location. The remaining tissue that was unable to be destroyed may continue to cause seizures. Some patients may have a secondary epilepsy, which means that they have seizures that arise from another part of the brain in addition to seizures arising from the HH. The MRI-guided laser ablation does create some minimal scar, but this scar does not cause seizures.

IF SEIZURES RETURN, WHAT CAN BE DONE?

If a patient continues to have seizures 1 year after MRI-guided laser ablation surgery, their case is thoroughly reviewed by the epilepsy and neurosurgery teams. All options are considered including additional MRI-guided laser ablation surgery, stereotactic radiosurgery, and endoscopic resection.

WHAT IF A PATIENT HAS OTHER SEIZURE TYPES BESIDES GELASTIC SEIZURES?

Some patients have a secondary epilepsy, which means that they have seizures that arise from another part of their brain in addition to seizures arising from the HH. Laser ablation of the HH will not affect secondary epilepsy coming from elsewhere in the brain, but this doesn’t mean the secondary epilepsy cannot be treated. Each patient’s case is different. For patients that have a secondary epilepsy, a number of treatment options may be available including further surgical intervention, vagus nerve stimulation (VNS), responsive neurostimulation (RNS), medications, clinical trials, and dietary therapies.

HOW MANY MRI-GUIDED LASER ABLATION SURGERIES HAVE BEEN COMPLETED AT TEXAS CHILDREN’S ON PATIENTS WITH HH?

Texas Children’s Hospital was the first hospital in the world to use MRI-guided laser ablation surgery to destroy epilepsy-causing lesions within the brain. Over 74 MRI-guided laser ablation surgeries on 59 patients with HH have been completed at Texas Children’s as of March 2017. This technology has been used a total of over 140 times at Texas Children’s as of March 2017, including on patients with other types of epilepsy-causing lesions.

WHAT IS THE AGE RANGE OF PATIENTS THAT HAVE UNDERGONE MRI-GUIDED LASER ABLATION SURGERY AT TEXAS CHILDREN'S? IS THERE AN ADVANTAGE TO WAITING UNTIL PATIENTS ARE OLDER FOR SURGERY?

MRI-guided laser ablation surgery has been performed at Texas Children's on patients ranging from 5 months old into the mid-20s. The risks and potential benefits of each patient's case is discussed with their family. The surgery is technically more difficult in babies as the skull is still soft and not yet fused, but there are safe ways to fixate the head in very young children so laser ablation can be performed in a patient of any age. Seizures limit the ability of normal brain to form important connections, so there is a neurologic advantage to stopping seizures earlier in life.

WILL THIS SURGERY IMPROVE DEVELOPMENT AND BEHAVIOR?

Many patients with HH experience challenges with behavioral and psychiatric symptoms, including rage attacks, oppositional-defiant disorder, and ADHD. About 50% of patients that have undergone MRI-guided laser ablation surgery at Texas Children's report some improvement in behavior. Patients with HH may also have cognitive challenges. Stopping seizures may improve brain development, but many patients with HH continue to experience challenges with behavior and development after MRI-guided laser ablation surgery.

WHAT TYPE OF WOUNDS ARE TYPICAL AFTER MRI-GUIDED LASER ABLATION SURGERY?

Typically, each patient will have 1 stitch where each laser applicator was placed. These stitches are dissolvable. Young children will have similar incisions for each of the markers placed in the skull for catheter guidance. Patients should shower 48 hours after surgery and should have the wound examined by the surgeon 1 week after surgery. The patient's head will not be shaved for surgery.

ARE PATIENTS USUALLY IN PAIN AFTER MRI-GUIDED LASER ABLATION SURGERY?

The surgical approach involves multiple small scalp wounds (where the head frame was placed), which are not stitched closed, one or more small cranial openings (where the laser applicators were placed), which are stitched closed, or small incisions from the bone markers. Patients may have a mild headache post-operatively that can be controlled with oral medications. Patients typically leave the hospital on the first or second day after surgery with over-the-counter pain medication, antibiotics, and steroids.

WHEN DO PATIENTS NEED TO FOLLOW-UP AT TEXAS CHILDREN'S AFTER SURGERY?

In addition to regular post-surgical follow-up appointments, patients that have undergone MRI-guided laser ablation surgery should have an MRI scan and an appointment with a neurosurgeon and neurologist at both 3 months and 1 year after surgery. In patients who are coming from out of state or outside of the U.S., choosing 1 of these 2 times is typically sufficient. The Texas Children's team will assist with making arrangements and collaborating with patient's local neurology team for follow-up care.

WHAT ARE THE RISKS OF THE PROCEDURE?

No surgical procedure is without risks. The hypothalamus has an important role in the endocrine system and is located in the center of the brain near many critical structures. Any surgery to this part of the brain carries a chance of complications. Risks of MRI-guided laser ablation surgery include memory deficit, diabetes insipidus, hypothalamic obesity, and hemorrhage. When planning a patient's MRI-guided laser ablation, the surgeon places several safety markers around the critical brain structures to avoid damaging any tissue other than the HH. MRI-guided laser ablation surgery is safer and significantly less-invasive than open brain surgery with a craniotomy or endoscopic surgery.

HOW LONG DO PATIENTS TYPICALLY STAY AT TEXAS CHILDREN'S AFTER SURGERY?

Each patient and case is unique. Patients are typically discharged from the hospital the following day after MRI-guided laser ablation surgery. For those who are coming from out of state or outside of the U.S., plans should be made to stay in the Houston area for 1 week after hospital discharge until follow-up with the neurosurgeon has been completed. The patient will be cleared to travel home at that time.

SHOULD A PATIENT'S SEIZURE MEDICATIONS BE CONTINUED PRIOR TO SURGERY?

In general, patients should continue their regularly prescribed medications until surgery. The seizure medication Depakote® (valproate, divalproex sodium) can interfere with how the blood clots, so it is typically recommended that patients wean this medication at least several weeks prior to surgery. This can be done under the direction of the patient's local neurologist.

CAN A PATIENT'S SEIZURE MEDICATIONS BE DISCONTINUED AFTER SURGERY?

Texas Children's typically does not recommend any changes to a patient's seizure medications until 1 year after surgery unless there are profound side effects that need to be addressed. For patients that travel from out of state or outside of the U.S., seizure medications will be managed by the epilepsy team while at Texas Children's. Upon return home, a patient's local neurologist will resume managing medications. The Texas Children's epilepsy team collaborates closely with local neurologists to provide medication and treatment recommendations.

DOES TEXAS CHILDREN'S OFFER OTHER SURGICAL APPROACHES TO MANAGING HH, SUCH AS GAMMA KNIFE (STEREOTACTIC RADIOSURGERY) AND OPEN RESECTION/DISCONNECTION OF HH?

Texas Children's is one of the busiest comprehensive epilepsy centers in the world and offers radiosurgical, open, endoscopic and MRI-guided laser ablation treatments for HH and other neurosurgical conditions. In addition, Texas Children's has a robust neurostimulation program for the treatment of epilepsy. An interdisciplinary epilepsy conference is held weekly to decide the best treatment options for each patient. The outcome of this conference is discussed with each patient and their family.

ARE PATIENTS FROM OUTSIDE OF THE U.S. ACCEPTED?

Our international department is available to assist patients that live outside of the U.S. that are interested in traveling to Texas Children's for medical care. They will assist with all necessary paperwork, collection of medical records, and financial questions and clearance. If you live outside of the U.S., please contact our International Office at 832-824-1138 or international@texaschildrens.org to begin the referral process. Interpreters are available.

WHERE DO FAMILIES TYPICALLY STAY DURING THEIR TIME AT TEXAS CHILDREN'S?

If a patient is scheduled for an inpatient admission in the Epilepsy Monitoring Unit prior to surgery, there is a pull-out bed available within the room. All rooms in the EMU are private and a parent or guardian must stay with the patient at all times. Siblings are encouraged to visit but discouraged from staying overnight in the room. When patients are not scheduled for an inpatient admission prior to surgery, families often choose to stay in a hotel nearby the hospital. Please visit texaschildrens.org/patients-and-visitors for more information about planning for surgery at Texas Children's.

HOW MUCH DOES MRI-GUIDED LASER ABLATION SURGERY COST?

Texas Children's accepts a variety of insurance plans and has a billing department that can assist with financial options once it is determined that a patient is a candidate for MRI-guided laser ablation surgery. We encourage our patient families to contact their insurance providers to determine if surgery and testing at Texas Children's will be covered.

WHERE CAN I READ MORE ABOUT HH?

To learn more about hypothalamic hamartomas, please visit hopeforHH.org or rarediseases.org.