Acta Scientific Neurology (ASNE)

Volume 9 Issue 7 July 2026

Case Report

Robotic Stereotactic Radiofrequency Disconnection of Hypothalamic Hamartoma for Drug-Refractory Epilepsy: An Emerging Technique

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DOI: 10.31080/ASNE.2026.09.0921

Received: May 15, 2026

Published: June 30, 2026

Abstract

Hypothalamic hamartomas (HH) are rare, benign developmental malformations frequently associated with drug-refractory epilepsy, particularly gelastic seizures. Hypothalamic hamartomas attached to the hypothalamus, pose unique technical challenges for conventional surgical approaches due to deep-seated location and ill-defined hypothalamic interfaces. Robotic stereotactic radiofrequency (RF) disconnection offers submillimetric precision targeting and real-time intraoperative verification, representing an advanced minimally invasive alternative. We present a 3.5-year-old boy with 1.5 years of pharmacoresistant gelastic and focal seizures in whom MRI demonstrated a type 3B HH attached to the right hypothalamus. Presurgical evaluation including video-electroencephalography and endocrine profiling excluded significant hormone dysfunction except subclinical hypothyroidism. The planned robotic stereotactic RF disconnection utilizes CT-MRI fusion for trajectory planning, AutoGuide robot-assisted probe placement, and intraoperative O-arm CT confirmation, with 13 lesions through 4 trajectories at 74°C for 60 seconds per lesion. This case exemplifies the application of robot-assisted stereotactic RF disconnection for deep-seated, sessile intraventricular HH, balancing high seizure control rates with preservation of hypothalamic function and minimal permanent morbidity.

Keywords: Hypothalamic Hamartoma; Gelastic Seizures; Drug-Refractory Epilepsy; Robotic Stereotactic Radiofrequency Disconnection; Minimally Invasive Neurosurgery

Introduction

Hypothalamic hamartomas are rare, benign malformations arising from the tuber cinereum or mamillary bodies that are strongly associated with chronic pharmacoresistant epilepsy [1]. These lesions present a unique epileptic syndrome characterized by stereotyped gelastic (inappropriate laughter) and dacrystic (crying) seizures, often accompanied by focal motor events and behavioral arrest [1,2]. The hamartomatous tissue generates epileptiform activity that may rapidly propagate to distributed cortical networks, making seizure control dependent on effective disconnection of the hamartoma from its epileptogenic interface with the hypothalamus rather than on lesion volume reduction [3,4].

HH morphology is classified according to the Delalande-Fohlen system, with type 3 lesions being sessile and type 3B specifically referring to completely intraventricular, sessile hamartomas attached to the hypothalamus [5]. These deep-seated lesions with ill-defined interface anatomy present significant technical difficulty for open microsurgical resection and endoscopic disconnection, as direct visualization is limited and the risk of hypothalamic morbidity (including endocrine dysfunction, memory impairment, and autonomous behavioral changes) is substantial [5,6].

Recent advances in minimally invasive neurosurgery have established robotic stereotactic radiofrequency (RF) ablation as a viable and increasingly preferred approach for deeply situated HH, particularly when sessile intraventricular morphology precludes conventional open or endoscopic techniques [7,8]. Robotic systems integrating CT-MRI fusion, bone fiducial registration, and intraoperative imaging verification enable submillimetric trajectory accuracy and controlled thermal lesioning at the hamartoma-hypothalamus interface with minimal collateral tissue injury [7-9].

This report describes the presurgical evaluation, clinical presentation, neuroimaging findings, and planned robotic stereotactic RF disconnection in a pediatric patient with drug-refractory HH epilepsy and type 3B lesion morphology.

Case Presentation

Clinical history and perinatal background

A 3.5-year-old male child was brought for neurosurgical evaluation with a 1.5-year history of recurrent drug-refractory seizures occurring at a frequency of 1–2 episodes per day. There was no significant perinatal insult, and developmental milestones were age-appropriate. Prior to evaluation, the patient had been treated with valproate and clobazam without adequate seizure control.

Seizure semiology

The predominant seizure phenotype consisted of brief, stereotyped episodes of inappropriate laughter and crying, consistent with Gelastic and dacrystic seizures. These events were sudden in onset, behaviorally incongruent, and not associated with clear external emotional triggers. In addition, focal seizures characterized by left facial deviation, sometimes followed by postictal sleep, were documented. This semiology is highly characteristic of HH-related epilepsy and represents the most specific clinical marker for this disorder.

Neurological examination

General neurological examination revealed no focal motor, sensory, cerebellar, or pyramidal deficits. There were no signs of raised intracranial pressure and no clinical evidence of precocious puberty. Neuropsychological assessment was within the average range for age-matched peers, and no overt behavioral disturbance was documented at the time of evaluation.

Endocrine profiling

A comprehensive endocrine panel was obtained to exclude central precocious puberty and other hypothalamic endocrine dysfunction. Serum thyroid-stimulating hormone (TSH) was mildly elevated (6.8 mIU/L; normal 0.5–4.5 for age), with normal free thyroxine (T4) (11.2 pmol/L; normal 10–19 for age), consistent with subclinical hypothyroidism. Morning cortisol was within normal range (485 nmol/L; normal 200–700), and prolactin was age-appropriate (8.4 mIU/L; normal 5–25 for prepubertal children). Luteinizing hormone (LH) and follicle-stimulating hormone (FSH) were in the prepubertal range (LH 0.3 IU/L, FSH 0.5 IU/L), and testosterone was appropriately low for a prepubertal male (0.35 nmol/L; prepubertal <2 nmol/L). This profile excluded biochemical evidence of central endocrine hyperfunction while documenting mild thyroid dysfunction requiring monitoring.

Video-Electroencephalography (Video-EEG) findings

Prolonged video-EEG monitoring was performed to characterize the electroclinical events and assess localization. EEG findings were nonspecific and poorly localizing, demonstrating generalized background attenuation, diffuse rhythmic theta activity, and multifocal low-amplitude epileptiform discharges without a clear focal onset zone. This pattern is typical of deep hypothalamic generators and reflects rapid intrahemispheric propagation of ictal activity. Interictal periods showed mildly disorganized background activity but no focal abnormalities. The discordance between clinically stereotyped semiology (suggesting focal origin) and diffuse EEG findings is characteristic of HH epilepsy and reflects the location of the primary generator deep within the midline hypothalamus [2].

Magnetic resonance imaging findings

High-resolution MRI (HARNESS protocol) of the brain was performed using 3-Tesla imaging. The study revealed a type 3B hypothalamic hamartoma that was completely intraventricular, sessile, and attached to the right hypothalamus.

On T1-weighted images, the lesion was isointense to adjacent gray matter. On T2-weighted images, the hamartoma was mildly hyperintense relative to gray matter, with a well-defined, non-infiltrative margin (Figure 1- A,B and C). Post-contrast T1-weighted imaging demonstrated no significant enhancement of the lesion, which is typical for hamartomatous tissue composed of gray matter without abnormal vasculature.

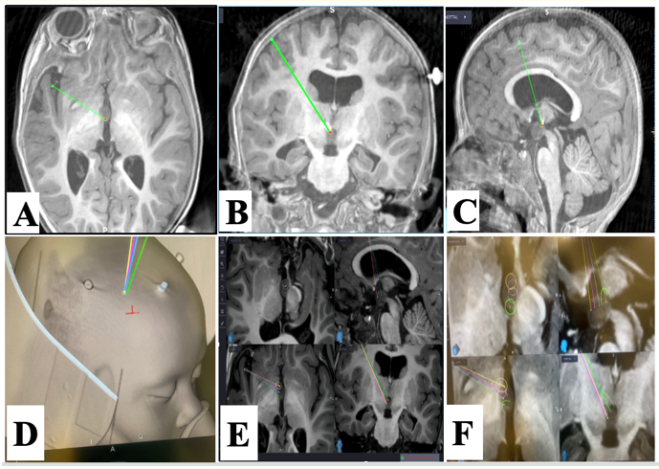

Figure 1: A,B,C) Show axial, coronal and sagittal T1weighted MRI images with type 3b type hypothalamic hamartoma. D) Shows entry point for radiofrequency probe in pre-coronal area. Note that multiple trajectories were planned in the same twist drill craniostomy. E and F) Show planning of trajectories, interface between hamartoma and hypothalamus and extent of lesions.

Neuropsychological assessment

Formal neuropsychological evaluation demonstrated average overall intellectual function appropriate for age. No significant deficits in attention, language, or visuospatial processing were identified, and behavioral screening was unremarkable.

Surgical planning and rationale

- **Surgical options:** Three principal surgical approaches were considered: endoscopic disconnection, open microsurgical resection with transcallosal approach, and minimally invasive stereotactic radiofrequency disconnection with robotic guidance.
- **Open Microsurgical Resection:** Traditional open approaches via transcallosal or subtemporal trajectory provide direct visualization but require wide dural openings, brain retraction, and direct hypothalamic manipulation, leading to higher rates of permanent endocrine dysfunction, memory disturbance, and hemiparesis in reported series.
- **Endoscopic Disconnection:** While endoscopic techniques offer minimized invasiveness, visualization of the sessile intraventricular attachment is frequently obscured by ventricular crowding, and the narrow operative corridor increases risk of incomplete disconnection and injury to adjacent deep venous structures or hypothalamic tissue. The completely intraventricular sessile morphology created a narrow surgical corridor and rendered the hypothalamic-hamartoma interface difficult to visualize directly.
- **Robotic Stereotactic Radiofrequency Disconnection:** This minimally invasive option integrates advanced neuronavigation, submillimetric stereotactic accuracy, and real-time intraoperative imaging confirmation, enabling targeted thermal lesioning at the hamartoma-hypothalamus interface while minimizing collateral tissue injury.

Rationale for robotic stereotactic RF disconnection

Given the completely intraventricular, sessile morphology of the type 3B lesion with attachment to the right hypothalamus, robotic stereotactic RF disconnection was selected as the optimal approach. The fundamental principle underlying this choice is that seizure control in HH epilepsy depends on effective disconnection of the hamartoma from the hypothalamic epileptogenic network rather than on gross total volumetric removal [3,4]. Robotic systems enable multiple non-intersecting trajectories to be planned through three-dimensional CT-MRI fusion, allowing comprehensive disconnection of the lesion while avoiding direct manipulation of hypothalamic tissue and preserving critical anatomical structures (foramen of Monro, fornices, deep veins, mamillary bodies) [7-9].

Planned Procedure: Robotic Stereotactic radiofrequency disconnection

Operative technique

The procedure was performed under general Anesthesia. The head was fixed in a three-pin skull clamp (Mayfield), and bone fiducials were surgically placed at predefined locations on the skull for stereotactic registration.

- **CT-MRI Fusion and Trajectory Planning:** High-resolution CT imaging was obtained with fiducials in place. This CT dataset was registered to the preoperative high-resolution MRI using rigid-body alignment algorithms. Multi-planar three-dimensional trajectory planning was performed to optimize access angles while avoiding blood vessels, sulci, ventricles etc. (Figure 1).
- **Robotic-Assisted Probe Placement:** The AutoGuide robotic system was registered to the stereotactic frame using fiducial-based localization. The radiofrequency probe will be mounted on the robotic arm and guided along the planned trajectories.
- **Intraoperative O-arm CT Confirmation:** After probe placement along first trajectory, intraoperative cone-beam CT imaging was acquired to confirm precise positioning before lesioning. This real-time verification step eliminates reliance on indirect anatomical landmarks and ensures quantitative accuracy (Figure 2). This step eliminated errors arising from CT-MRI fusion, trajectory alignment and brain shift.
- **Radiofrequency Lesioning:** Thermal lesioning was performed using a temperature-controlled radiofrequency generator. The protocol includes creation of 13 lesions through 4 trajectories, each delivered at 74°C for 60 seconds. This temperature and duration have been selected based on prior series to achieve reliable tissue necrosis sufficient for effective disconnection while minimizing thermal spread beyond the target interface.
- **Closure and Recovery:** After completion of lesioning and confirmation that all trajectories were completed, the probe was withdrawn, fiducials were removed, and the scalp was closed in standard fashion. The patient was extubated and transferred to the intensive care unit for postoperative monitoring.
- **Follow up :** Duration of follow up was 2.3 years. The child, now, goes to normal school with normal neurocognitive development. He is on thyroxine supplement. Motor and cognitive milestones are appropriate for the age. Two episodes of aura were reported since the time of surgery (ILAE 2 category). He is under regular follow up with surgical epilepsy team and paediatric endocrinologist.

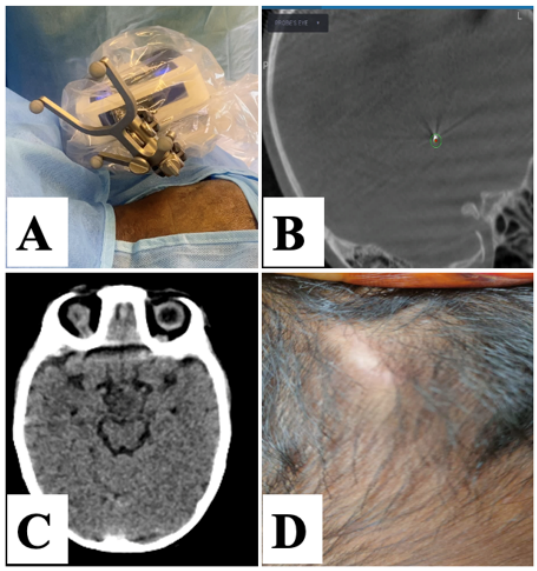

Figure 2: A) Robotic arm (Autoguide, Medtronic) is positioned in planned trajectory. B) Confirmation accuracy of probe placement using intra-operative CT. Red dot denotes planned trajectory, and white dot denotes actual trajectory (RF Probe). C) Shows hypodensity in hypothalamus indication lesioning. D) Shows extent of surgical scar after healing.

Literature Review and Discussion

Pathophysiologic basis for disconnection strategy

Recent electrophysiologic and imaging studies have clarified that HH epilepsy arises from intrinsic hyperexcitability within the hamartomatous tissue itself, not from abnormal structural compression of the hypothalamus [3,4]. Seizures are generated by the hamartoma and propagate rapidly to distributed cortical networks via the hypothalamic-thalamic-cortical system. Effective seizure control is achieved by disruption of this propagation pathway (i.e., disconnection) rather than by complete resection of the hamartoma volume [3,4]. This pathophysiologic principle

validates the RF disconnection strategy of creating multiple lesions at the hamartoma-hypothalamus interface to block propagation, as opposed to attempted gross total removal which carries greater morbidity and does not necessarily improve outcomes.

Overview of surgical approaches for hypothalamic hamartoma epilepsy

The literature demonstrates that seizure control outcomes in HH epilepsy depend primarily on effective disconnection of the hamartoma from the hypothalamic epileptogenic focus rather than on the extent of lesion resection [3,4]. Comparison of different surgical modalities reveals important distinctions in seizure freedom rates, morbidity profiles, and technical feasibility (Table 1).

Author (s)/Surgical Approach	Year	No. patients	Surgical approach	Seizure freedom %	Complications
Stereotactic RF Ablation					
Reinacher., *et al*. [10]	2025	35	Stereotactic RF thermo-coagulation	60 (12 mo)	14% mild hypothalamic dysfunction
Rampp., *et al*. [11]	2024	150	Stereotactic RF ablation	73	Low; weight gain, hormonal changes
Shirozu., *et al*. [12]	2021	131	Stereotactic RF thermo-coagulation	88.6 (gelastic)	Hormonal (12%), weight gain (7.5%)
Li., *et al*. [13]	2020	20	Repeat stereotactic RF	55 (gelastic-free)	Transient; low permanent
Ramesh D., *et al*. (14)	2020	16	Stereotactic RF ablation	75	One – twice RF Ablation Three- thrice RF Ablation
Franzini., *et al*. [15]	2016	25	Stereotactic RF thermo-coagulation	68	Minimal; hematoma rare
Parrent [16]	2004	13	Stereotactic RF ablation	62	Transient memory (15%)
Laser Ablation (LITT)					
Nariai., *et al*. [17]	2021	25	MR-guided LITT	64	Transient deficit (12%); permanent 4%
Pati., *et al*. [18]	2019	19	Robotic LITT (ROSA)	68	Infection (5%); hemorrhage (5%)
Endoscopic Disconnection					
Koutromanou., *et al*. [19]	2024	45	Endoscopic disconnection	58	CSF leak (11%); DI (7%)
Procopio., *et al*. [20]	2022	24	Endoscopic hamartomotomy	62	Hypothalamic obesity (17%)
Open Microsurgical					
Ng., *et al*. [21]	2019	72	Transcallosal/interhemispheric	65	DI (22%); memory deficit (18%)

Table 1: Comprehensive Literature Review Table (2000–2026). Literature review comparing various surgical techniques and their results.

Discussion

Open microsurgical resection: Morbidity profile

Open microsurgical approaches via transcallosal or subtemporal trajectories provide wide operative exposure and direct visualization of the lesion-hypothalamus interface. However, achievement of this exposure requires wide dural opening, significant brain retraction, and direct manipulation and dissection of hypothalamic tissue. Reported seizure freedom rates of 55–72% are achieved [21]. Large series document permanent endocrine dysfunction in 22–30% of patients, permanent memory deficits in up to 18%, and permanent hemiparesis in 12% [21]. Operative mortality, while rare, has been reported at 3% in the largest series. These complications reflect direct thermal and mechanical injury to hypothalamic tissue and adjacent eloquent structures inherent to open resection.

Endoscopic approaches: Limitations and outcomes

Endoscopic disconnection offers advantages of minimal invasiveness and direct visualization. However, in deeply situated intraventricular sessile lesions, visualization of the hamartoma-hypothalamus interface is frequently obscured by ventricular crowding, cerebrospinal fluid pulsation, and the narrow operative corridor created by third ventricular anatomy [19,20]. Large endoscopic series report seizure freedom rates of 58–62%, and transient diabetes insipidus (7%) that reflect direct hypothalamic manipulation [20].

Stereotactic RF disconnection: Technical advantages and clinical outcomes

Stereotactic RF Disconnection (frame based/frameless) integrate several technological advantages such as submillimetre accuracy, better visualisation of hamartoma-hypothalamus interface while planning, and intraoperative confirmation of probe and repeatability without cumulative morbidity. For this patient's type 3B completely intraventricular, sessile hamartoma attached to the right hypothalamus, robotic RF disconnection is particularly well suited. The completely intraventricular location precludes easy access via transcortical or transventricular endoscopic corridors without significant brain or ventricular structure manipulation. The sessile attachment means the lesion has no stalk and is broadly based on the hypothalamus, making sharp dissection of an intact interface difficult or impossible via endoscopy. The deep midline location and intimate relationship to the hypothalamus make open transcallosal approaches high-risk for hypothalamic morbidity. In contrast, robotic RF disconnection can create multiple small lesions at the periphery of the lesion, effectively isolating it from the surrounding hypothalamic network while minimizing injury to hypothalamic tissue itself.

Large multicenter series of stereotactic RF ablation report seizure freedom rates of 60–89%, with gelastic seizures showing particularly high control rates (88.6% in the largest series of 131 patients) [10-16]. Permanent major complications are uncommon; hormonal changes occur in 7–14% (predominantly reversible weight gain), and permanent endocrine dysfunction is rare (<2% in most series) [10-12]. Memory deficits and hemiparesis are either absent or transient in RF series [10-12].

Conclusion

This case exemplifies the evolving paradigm in HH epilepsy surgery toward minimally invasive, precision-guided techniques that prioritize safety and functional preservation alongside seizure control. For type 3B intraventricular sessile hamartomas, robotic stereotactic radiofrequency disconnection represents a rational, technically feasible, and evidence-supported approach that achieves seizure freedom rates while maintaining a low morbidity profile.

The planned procedure demonstrates integration of modern neurosurgical technology (robotic guidance, CT-MRI fusion, real-time intraoperative imaging) with established principles of epilepsy surgery (functional disconnection rather than gross total removal) to optimize outcomes in a challenging lesion type. Ongoing prospective data collection and long-term follow-up remain essential to refine patient selection, predict individual outcomes.

Funding

No specific grants from any funding agency in the public, commercial, or not-for-profit sectors.

Conflicts of Interest

The authors declare no conflicts of interest.

Data Availability

All data supporting this case report are contained within the manuscript. Additional clinical details are available upon request from the corresponding author.

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