



Posterolateral Disconnection as an Effective Technique in Drug Recalcitrant Epilepsy – A Case Report and Review of Literature

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Abstract

Refractory epilepsy surgeries have evolved over the years from resection to disconnection procedures reducing the perioperative morbidities and long-term complications, while yielding good outcomes. Posterior quadrant disconnection surgery accounts for less than 5% of overall epilepsy surgeries and is less frequently described in literature. We hereby report a case report with refractory epilepsy that underwent posterior quadrant disconnection and improved well. A quick review of literature is also supplied.

Keywords: Posterior quadrant disconnection; Medically recalcitrant epilepsy; Semiology

Introduction

Posterior quadrant disconnection is effective surgical procedure for medically refractory epilepsy arising from the posterior quadrant in carefully selected patients without morbidity or functional disability across various age groups especially in children.

Case Presentation

The patient was a 9 years old male child. He was admitted into or care with drug recalcitrant epilepsy. Had seizures from the first week after birth. He was on medications for next 5 years. Seizures occurred despite the intense medications he used. The seizure semiology was as follows. No aura occurred, but the child would slowly slump to one side, eyes to moving to the left with left hand automatisms. There would be absolutely no movements on the right side. The seizure would last from 3 min to 4 min at a time. The postictal phenomena consisted of headache and drowsiness. The frequency was once every day. No nocturnal events would occur. The child was the first born of a non-consanguineous parentage and born without incident at full term by a normal delivery. He had neonatal jaundice and delayed cry at birth. As his growth progressed along with the seizures, his motor and speech milestones were mildly delayed. There was no family history of epilepsy. Now his seizures have increased for the past 2 years. He has been on multiple medications in the past, but with no seizure free periods. He is currently on the maximum dose per weight of sodium valproate, oxcarbazepine, clobazam and lamotrigine, with still no relief from his seizures. On examination, there was right hemianopia with mild spasticity in the right upper and lower limbs. Dexterity was difficult in the right hand. The child was left hander, the left side was normal. A detailed neuropsychological assessment showed significant cognitive issues which will be discussed later. He was subjected to the seizure identification and analysis battery of our institution. An MRI of the brain was done along with a PET CT and functional MRI to discern the damage in the brain. The MRI showed Gliotic lesions in the left periventricular region with extensive gliosis in the left temporal (particularly in the posterior neocortex), and parieto-occipital regions (Figure 1). The PET showed hypo metabolism over the left temporal, parietal and occipital lobes along with the left thalamus as well (Figure 2). An fMRI confirmed the hypo metabolism of the gliotic regions with the right sided posterior temporal region lighting up primarily for language stimulation. In addition to imaging studies electrophysiological assessment was also done with a video Electroencephalogram (EEG) showing frequent spikes over the left occipital and temporal regions with a generalized spike across the left hemisphere (Figure 3). A detailed neuropsychological assessment showed no memory deficits, but aggressive behavior issues with agitation. He was also shown to have below par scholastic performance. Language assessment showed a difficulty in speaking long sentences, but with otherwise good comprehension and fluency. Lastly an eye examination with visual fields, showed

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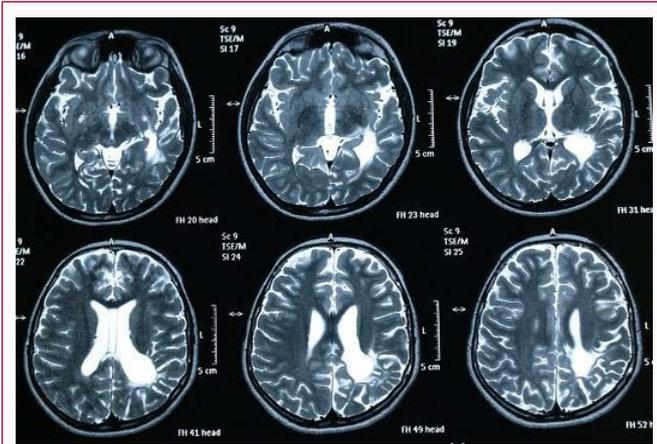


Figure 1: MRI of the brain in axial cuts showing extensive gliosis over the posterior aspect of the left side of the brain.

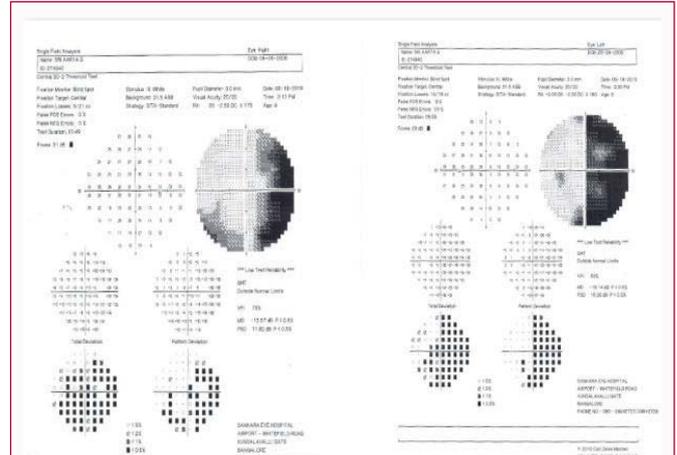


Figure 4: Perimetry showing hemianopia.

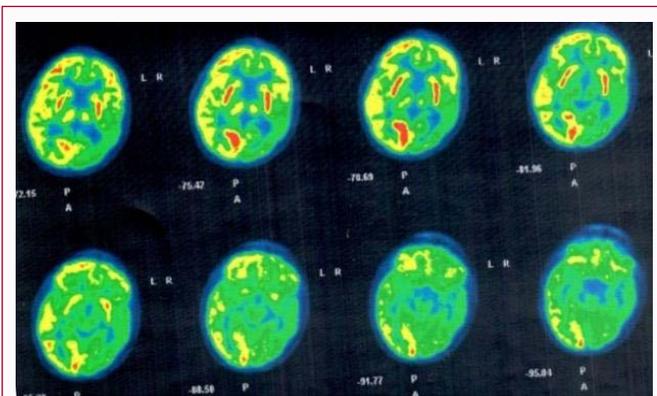


Figure 2: PET of the brain showing gross hypo-metabolism over the gliotic area.

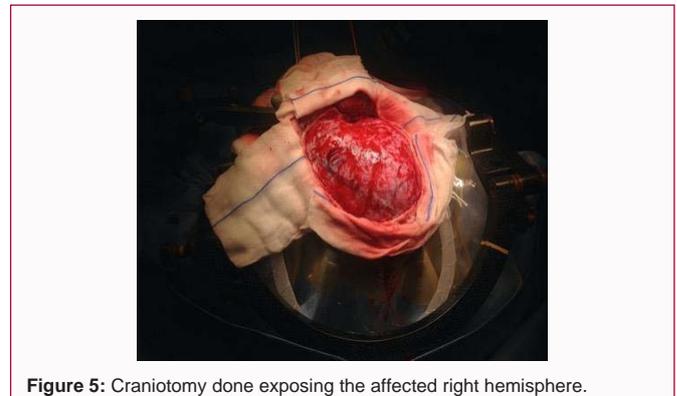


Figure 5: Craniotomy done exposing the affected right hemisphere.

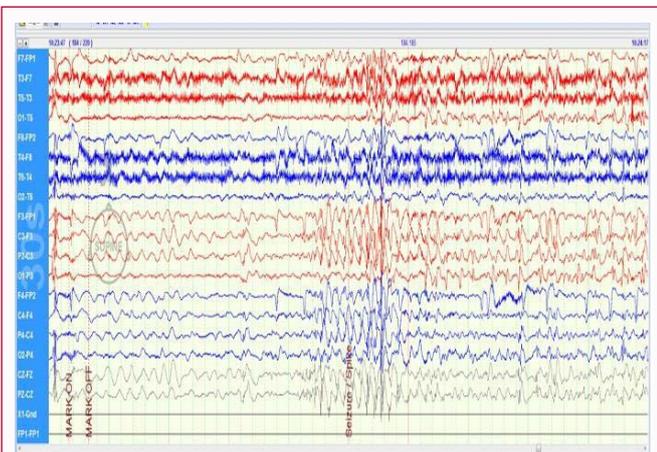


Figure 3: EEG of the patient showing epileptic form discharges localized to the affected area.

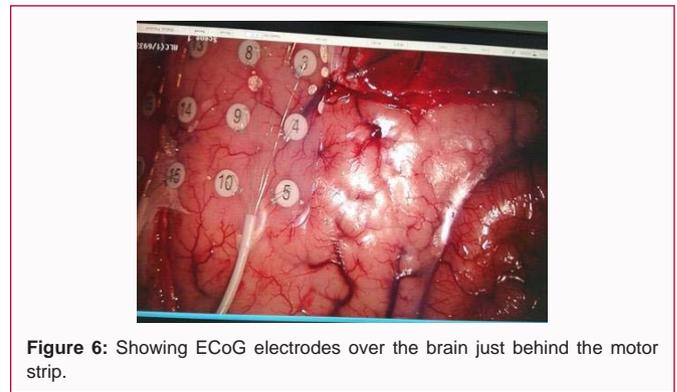
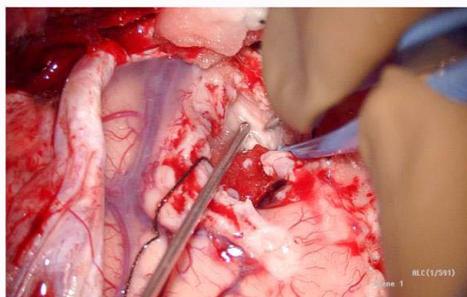


Figure 6: Showing ECoG electrodes over the brain just behind the motor strip.

an expected right hemianopia (Figure 4). As there was excellent clinic-radiological and electrophysiological concordance, the child was considered for posterolateral disconnection. The thought process regarding the choice of procedure amongst the options available is mentioned later. The child and his parents were counseled in detail regarding the nature of the disease, the need for surgery as especially regarding post-operative deficits such as hemiplegic (which may be permanent), speech loss (global aphasia), Hemianopia and persistent

seizures despite the procedure requiring lifelong anticonvulsants to be taken, were discussed. A pre op check up by the anesthetists was done and the child was wheeled into the OT. Surgery was performed under electrocorticographic guidance requiring no GABAergic medications to be given during the stimulation period. The child was put on a dexmedetomidine infusion to maintain anesthesia with an infusion of opioids for analgesia. The head was positioned on a sugita head frame, with the head turned to the right completely allowing a large craniotomy to be made, exposing the entire left hemisphere for surgery (Figure 5). Electrocorticographic (ECoG) was done and the margin between damaged and normal brain was delineated. The Motor strip was delineated as well, and the resection kept away from both the sensory and motor areas as much as possible (Figure 6). As per convention, the first step of the surgery after ECoG was



Video photograph 1: Showing Mesial temporal lobectomy.

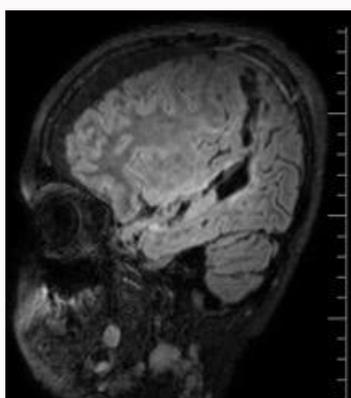


Figure 7: Showing the post op MRI of the brain. The disconnection is clearly visible.

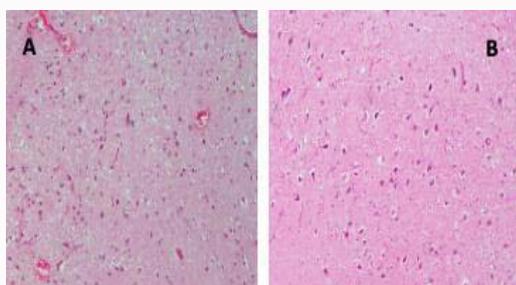


Figure 8: Paraffin section showing (A) architectural dyslamination, loss of neurons, (B) ischemic neurons.

a temporal lobectomy. The temporal neocortex was removed as per convention up to 5 cm from the temporal pole. Then the mesial temporal lobe was removed (Video photograph 1). The next stage was to follow the ventricle up to the ECoG margin delineated before. The parietal disconnection was donning first up to the falx. The inferior parietal/superior temporal disconnection was last and was done along connecting the 2 ends of the disconnection along the temporal horn. The last stage involved disconnecting the splenium from the parieto-occipital lobe. This ‘Intraventricular stage’ was done last through the atrium and body of the lateral ventricle which was exposed through the dissection. The fornix was exposed along the medial wall of the atrium and disconnected last completing the procedure. Post op, the child was extubated on table and was monitored in the ICU. Antiepileptics were maintained post-surgery. The drain was removed on POD3 and he was discharged after 5 days in hospital with no further seizures. His post op deficits included transient right foot and hand weakness and persistent hemianopia.

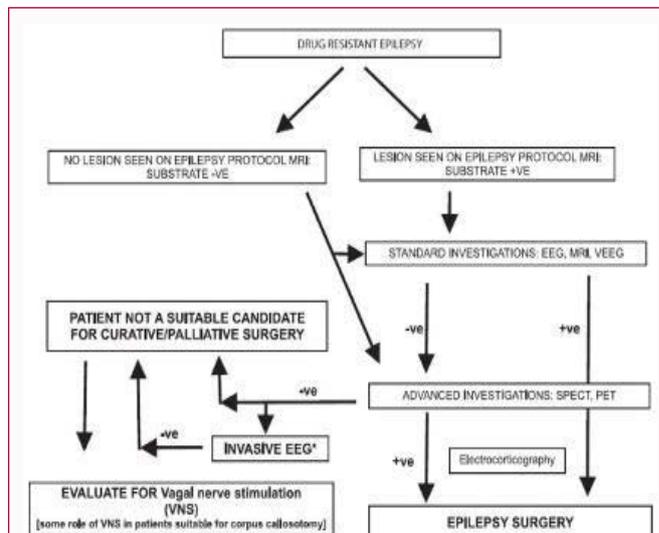


Figure 9: Flow chart showing a management paradigm for patients being subjected to epilepsy surgery.



Video photograph 2: Showing post operative recovery after 2 years of follow up.

No worsening of speech occurred. The weakness improved well after physiotherapy and after 3 months on review only a mild weakness of hand grip was present. His anticonvulsant medication was also scaled down to only sodium valproate on a minimal dosage with plans to reduce that as well. (Engel 1A outcome) After 2 years on follow up, he was seen walking without support with minimal weakness of the right upper limb alone (video photograph 2). His post op MRI suggested complete disconnection (Figure 7). The histopathological assessment of the resected hippocampus and amygdala showed architectural dyslamination, loss of neurons and ischemic neurons consistent with the gliosis and birth injury sustained by the child (Figure 8).

Discussion

Medically retractable epilepsy is often the source of discord between the students of different schools of epilepsy. The generally accepted criteria to consider surgery however are beyond reproach. Either a change in the semiology of the seizure pattern or the rule of 2 which consists of >seizures per week, >2 AEDs on full

dose, 2 neurologists agreeing upon the diagnosis, and >2 years of intractability, imply the need for surgical intervention. In diagnosing the type of surgery and the need for surgery, many protocols have been produced. We followed that proposed by Chandra et al. which is shown in Figure 9. The need for surgery thus clearly established, we proceeded to localize the lesion and propose surgery. Our choices were between resection and disconnection. As discussed by Jehi et al. suggest that resections have a greater incidence of failure and recurrence 56% as compared to 31% in disconnections [1-6]. Nooraine et al. further went to say that incidence of complications less in disconnections, and Immediate post-operative seizures were less in disconnections although they borneo long term bearing on prognosis (Jehi et al.). The problems with resection included the fact that there was diffuse gliosis without a well circumscribed lesion with multilobar involvement. The procedure would involve extensive surgery with significant blood loss, especially for a child, and a higher incidence of post-operative deficits and damage to the working brain. There was also a greater risk of scar epilepsy as well. Procedure wise, we followed the peri-insular posterior quadrant disconnection which is divided into 5 phases, starting with a temporal lobectomy both (neocortex and mesial lobe) followed by the parieto-occipital disconnection [7-10]. The stages included:

Temporal stage

The superior temporal gyrus is resected from temporal pole to posterior end of Sylvian fissure. The white matter of temporal stem is reached below the inferior margin of the insula. The temporal horn is opened along its entire length. The incision in superior temporal gyrus is continued from the temporal operculum to the parietal operculum and entered into the ventricle to display the tail of hippocampus and fornix in the medial wall of the atrium. The amygdala is then resected in the roof of the temporal horn till the optic tract is visualized. The connection with the head of hippocampus is sectioned.

Intraparietal disconnection

The motor cortex is identified by intraoperative stimulation. The parieto-occipital lobe disconnection is done from lateral to medially till the falx cerebri is encountered and from superior sagittal sinus superiorly to the parietal operculum behind the sensory strip inferiorly.

Intraventricular stage

The splenium is identified in the atrium at the junction of roof and medial wall of the lateral ventricle. The fibres originating from the parieto-occipital lobe and entering the splenium are sectioned. This incision is extended on the medial wall of the atrium to reach the floor and fornix along with the choroidal fissure. The fornix is sectioned to complete the disconnection. The arteries and veins over the cortex are preserved as far as possible. A search of recent literature showed most patients achieve Engel 1 and 2 by 2 years without resorting to further anticonvulsant therapy or further intervention [11,12]. There is a slight deterioration after 1 year which can be attributed to damage to the working brain (either with surgery or ischemia) leading to epilepsy to persist. Despite this, results for this procedure are excellent showing impressive seizure control and minimal deficits (Table 1). Few studies have evaluated surgical outcomes in posterior cortex epilepsies [13]. Boesebeck et al., reported 68.5 and 48% Engel class I postoperative outcome at 1 and 2 years, respectively [14]. Good prognostic indicators included, lateralizing auras, lateralizing clinical seizures or combination, tumoral aetiology, and absence of epileptiform discharges in the postoperative EEG. Jehi et al., in a

Table 1: Outcomes of the various studies on posterior quadrant disconnection in recent literature.

Study	Engel 1 (1 yr)	Engel 1 (2 yrs)
Boesebeck et al.	68.50%	48%
Jehi et al.	73.10%	65.80%
Daniel et al.	93%	84%
Lee YJ et al.	89%	75%
Rao et al.	98%	66.60%

similar study reported Engel class I postoperative outcome as 73.1% at 6 months, 68.5% at 1 year, 65.8% between 2 and 5 years, and 54.8% at 6 years and beyond [15]. Parietal resections fared worse outcome than occipital or parieto-occipital resections (52% seizure freedom vs. 89 and 93%, respectively, at 5 years). Patients with tumoral aetiology or dysplasia fared better. Most recurrences (75%) in this series occurred within the first 6 postoperative months. In a large series of 16 children who underwent PQ disconnection for refractory seizures, nine children (56%) were seizure free and five children (31%) had 50% reduction in seizures at a mean follow-up of 52 months [2]. Improvement in neuropsychological functioning and development occurred in all pediatric patients. Seizure control improved electrophysiological environment, better cognitive stimulation either structured or otherwise and reduced medications likely aided this improvement.

Conclusion

Posterior quadrant disconnection is a very effective surgical procedure for medically refractory epilepsy arising from the posterior quadrant in carefully selected patients without morbidity or functional disability across age groups especially children.

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