

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

Sibhi Ganapathy* and Lingaraju T

Neurosurgeon and Spine Surgeon, Sakra World Hospital, Bangalore, India

***Corresponding Author:** Sibhi Ganapathy, Neurosurgeon and Spine Surgeon, Sakra World Hospital, Bangalore, India.

Received: May 26, 2020; **Published:** September 30, 2020

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. We present a child with medically refractory Epilepsy which hemiparesis and hemianopia. The imaging was suggestive of extensive posterior quadrant gliosis secondary to neonatal asphyxia. After successful seizure localisation, using phase 1 and 2 investigations, a posterior quadrant disconnection was successfully carried out without incident. The child recovered and is seizure free with no additional deficits. Posterior quadrant disconnection surgery accounts for less than 5% of overall epilepsy surgeries and is less frequently described in literature. Yet it has become more acceptable to neurosurgeons and patients alike over the years. A quick review of literature is also supplied.

Keywords: Posterior Quadrant Disconnection; Medically Recalcitrant Epilepsy

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. This work has been formulated in line with SCARE criteria [1-13].

Case Report

The patient was a 9 years old male child. He was admitted into or care with drug refractory epilepsy. Had seizures from the first week after birth. He was on medications for next 5 years. Seizures occurred despite the 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 - 4 minutes 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 in frequency for the past 2 years. No new semiology of seizures presented. 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, oxcarbamazepine, clobazam and lamotrigine, with still no relief from his seizures.

Citation: Sibhi Ganapathy and Lingaraju T . "Posterolateral Disconnection as an Effective Technique in Drug Refractory Epilepsy - A Case Report and Review of Literature". *EC Neurology* 12.10 (2020): 86-95.

On examination, there was right hemianopia with mild spasticity in the right upper and lower limbs. Dexterity was lost in the right hand. The child was left handed, 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 hypometabolism over the left temporal, parietal and occipital lobes along with the left thalamus as well (Figure 2). An fMRI confirmed the hypometabolism of the gliotic regions with the right sided posterior temporal region lighting up primarily for language stimulation.

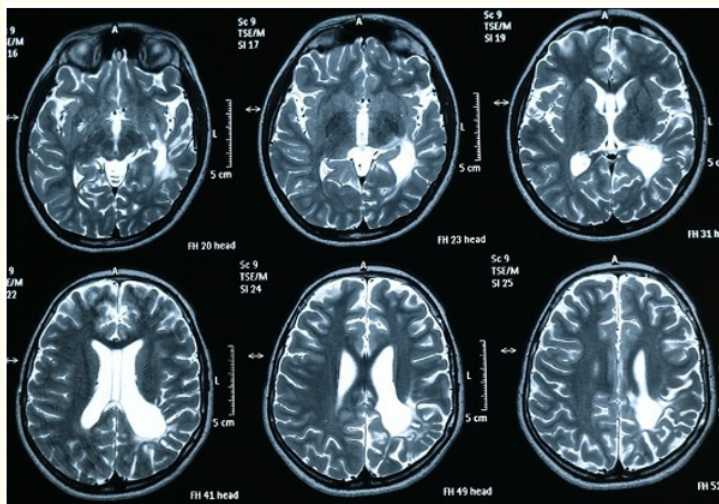


Figure 1: MRI of the brain in axial cuts showing extensive gliosis over the posterior aspect of the left side of the brain.

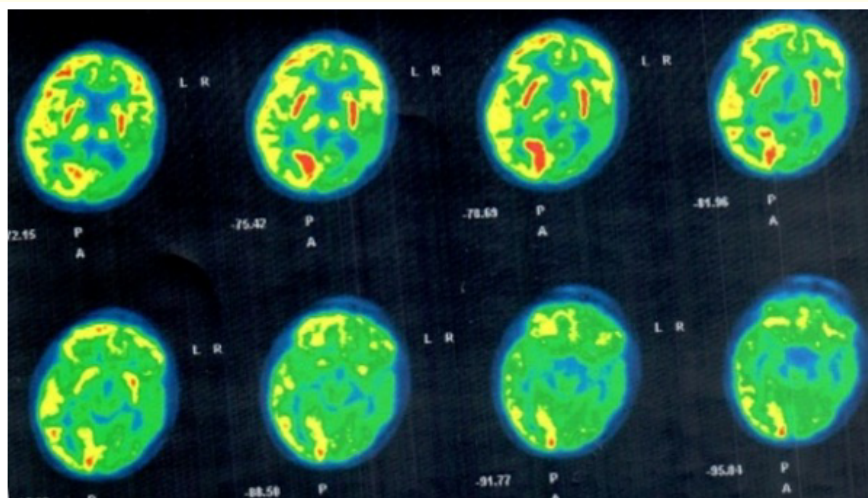


Figure 2: PET of the brain showing gross hypometabolism over the gliotic area.

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 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 an expected right hemianopia (Figure 4).

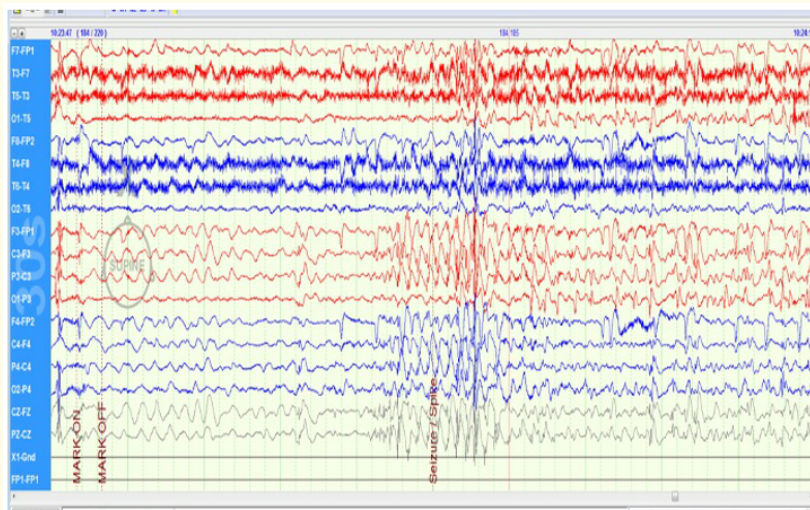


Figure 3: EEG of the patient showing epileptiform discharges localized to the affected area.

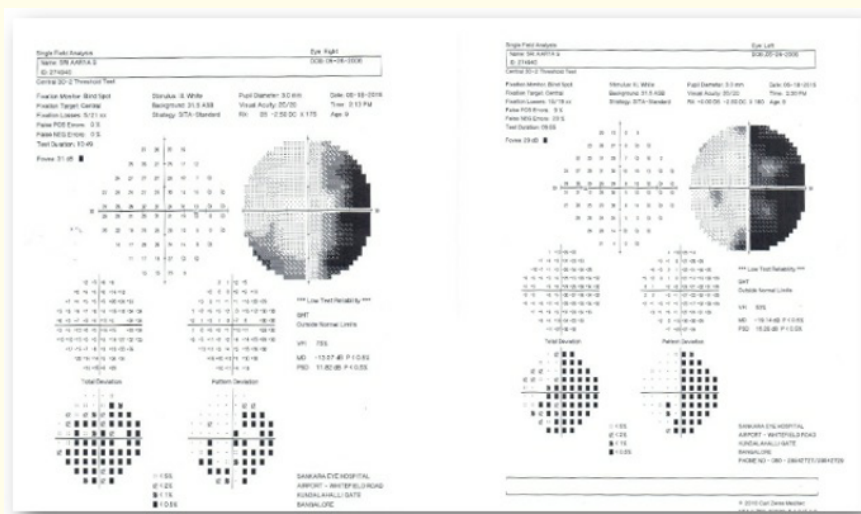


Figure 4: Perimetry showing hemianopia.

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 are mentioned later. The child and his parents were counselled in detail regarding the nature of the disease, the need for surgery along with probable post surgical morbidities.

Surgery was performed after all presurgical evaluation under electrocorticographic guidance requiring no GABAergic (Gama amino butyric acid) medications to be given during the stimulation period. The child was put on a dexmetromedine 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).

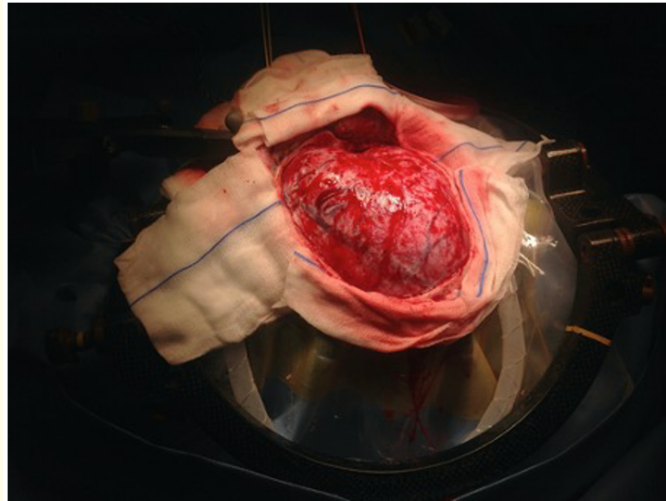


Figure 5: Craniotomy done exposing the affected right hemisphere.

Electrocorticography (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 a temporal lobectomy. The temporal neocortex was removed as per convention upto 5cm from the temporal pole. Then the mesial temporal lobe was removed (Video 1). The next stage was to follow the ventricle up to the ECoG margin delineated before. The parietal disconnection was don first upto 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.

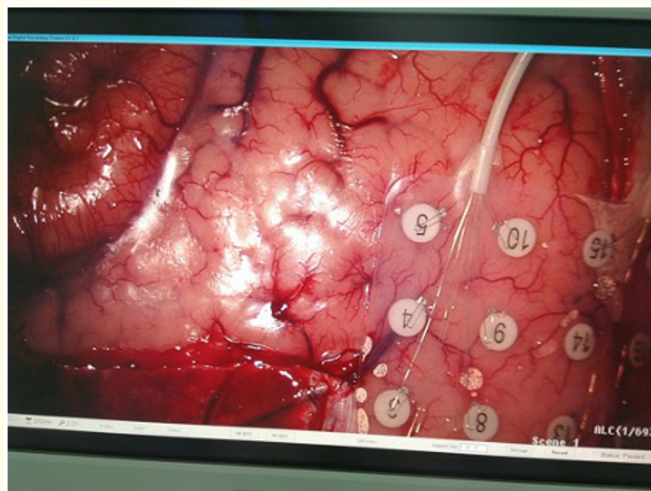
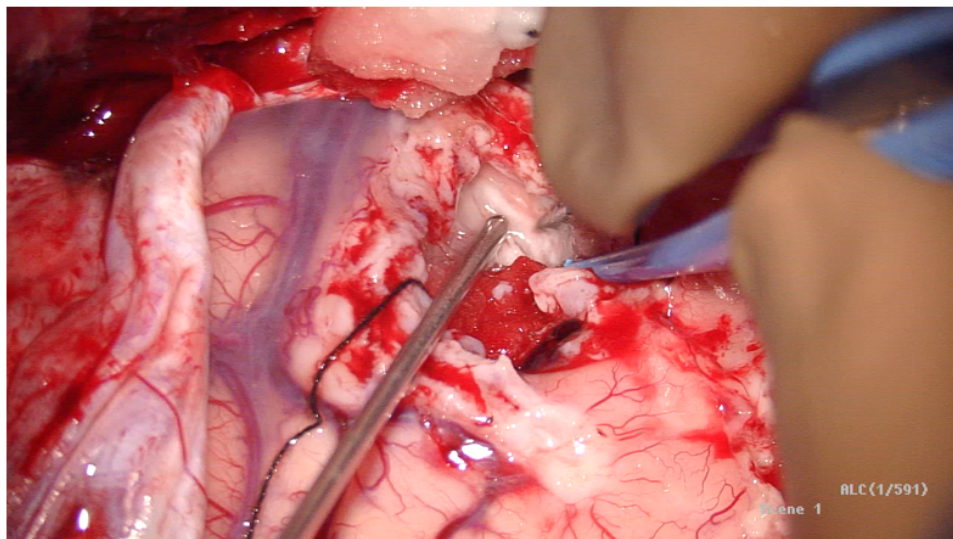


Figure 6: Showing ECoG electrodes over the brain just behind the motor strip.



Video 1: Showing mesial temporal lobectomy.

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 (post-operative day) 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. No worsening of speech occurred. The weakness improved well after physiotherapy and on review after 3 months 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 2**). His post op MRI suggested complete disconnection (Figure 7).

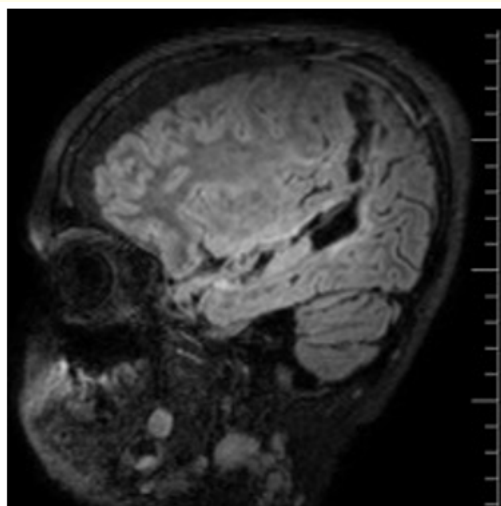


Figure 7: Showing the Post op MRI of the brain. The disconnection is clearly visible.



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

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).

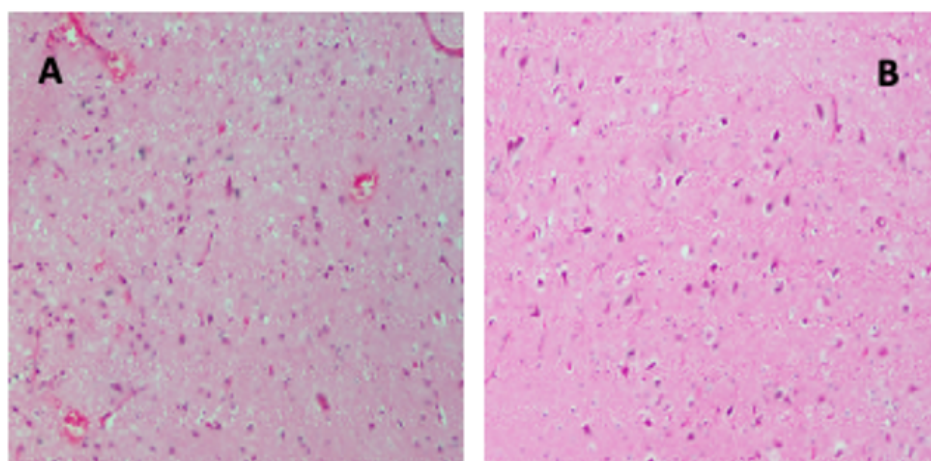


Figure 8: Paraffin showing (A) architectural dyslamination, loss of neurons, (B) ischemic neurons.

Discussion

Medically refractory epilepsy is often the source of discordance between the student 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 > 2 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 BS., *et al.* which is shown in figure 9.

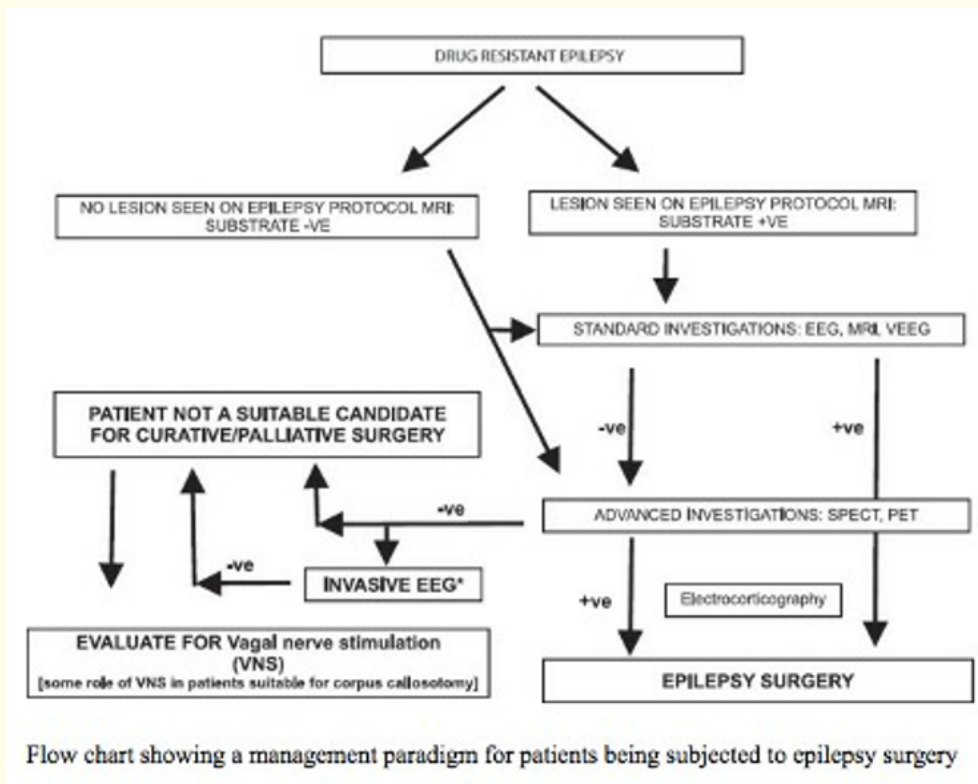


Figure 9: Flow chart showing a management paradigm for patients being subjected to epilepsy surgery.

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. Nooraine and associates further went to say that Incidence of complications less in disconnections, and Immediate post-operative seizures were less in disconnections although they bore no long term bearing on prognosis [15]. 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. 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. 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).

Study	Engel 1 (1yr)	Engel 1 (2yrs)
Boesebeck, <i>et al.</i>	68.5%	48%
Jehi, <i>et al.</i>	73.1%	65.8%
Daniel, <i>et al.</i>	93%	84%
Lee YJ, <i>et al.</i>	89%	75%
Rao, <i>et al.</i>	98%	66.6%

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

Few studies have evaluated surgical outcomes in posterior cortex epilepsies. 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 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 paediatric patients. Seizure control improved electrophysiological environment, better cognitive stimulation either structured or otherwise, and reduced medications likely aided this improvement [16].

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.

Informed Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Conflict of Interest

No conflict of interest present.

Funding

No financial grants used for the preparation and publication of this manuscript.

Bibliography

1. Rasmussen T. "Hemispherectomy for seizures revisited". *Canadian Journal of Neurological Sciences* 10 (1983): 71-78.
2. Mohamed AR., et al. "Temporoparietooccipital disconnection in children with intractable epilepsy". *Journal of Neurosurgery Pediatrics* 7 (2011): 660-670.
3. Källén K., et al. "Hypomotor seizures in infants and children". *Epilepsia* 43 (2002): 882-888.
4. Fogarasi A., et al. "A detailed analysis of symptomatic posterior cortex seizure semiology in children younger than seven years". *Epilepsia* 44 (2003): 89-96.
5. D'Agostino MD., et al. "Posterior quadrant dysplasia or hemi-hemimegalencephaly: A characteristic brain malformation". *Neurology* 62 (2004): 2214-2220.
6. Wyllie E., et al. "Successful surgery for epilepsy due to early brain lesions despite generalized EEG findings". *Neurology* 69 (2007): 389-397.
7. Novegno F., et al. "Epilepsy surgery of posterior quadrant dysplasia in the first year of life: Experience of a single centre with long term follow-up". *Seizure* 20 (2011): 27-33.
8. Lee YJ., et al. "Resective pediatric epilepsy surgery in Lennox-Gastaut syndrome". *Pediatrics* 125 (2010): e58-e66.
9. Kramer U., et al. "Focal features in West syndrome indicating candidacy for surgery". *Pediatric Neurology* 16 (1997): 213-217.
10. Chugani HT., et al. "Infantile spasms: I. PET identifies focal cortical dysgenesis in cryptogenic cases for surgical treatment". *Annals of Neurology* 27 (1990): 406-413.
11. Gupta A., et al. "Pediatric epilepsy surgery in focal lesions and generalized electroencephalogram abnormalities". *Pediatric Neurology* 37 (2007): 8-15.
12. Morrell F. "Varieties of human secondary epileptogenesis". *Journal of Clinical Neurophysiology* 6 (1989): 227-275.

13. Daniel RT, *et al.* "Posterior quadrant epilepsy surgery: Technical variants, surgical anatomy, and case series". *Epilepsia* 48 (2007): 1429-1437.
14. Boesebeck F, *et al.* "Lateralizing semiology predicts the seizure outcome after epilepsy surgery in the posterior cortex". *Brain* 125 (2002): 2320-2331.
15. Jehi LE, *et al.* "A longitudinal study of surgical outcome and its determinants following surgery". *Epilepsia* 50 (2009): 2040-2052.
16. Agha RA, *et al.* "For the SCARE Group. The SCARE 2018 Statement: Updating Consensus Surgical Case Report (SCARE) Guidelines". *International Journal of Surgery* 60 (2018): 132-136.

Volume 12 Issue 10 October 2020

©All rights reserved by Sibhi Ganapathy and Lingaraju T .