Introduction

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.[1] Posterior quadrant disconnection surgery accounts for less than 5% of overall epilepsy surgeries and is less frequently described in literature.[2] We hereby report a case series of seven consecutive patients with refractory epilepsy who underwent posterior quadrant disconnection.

Materials and Methods

Seven consecutive patients of refractory epilepsy who underwent posterior quadrant disconnection from a level four comprehensive epilepsy program over the last 3 years were reviewed.

Results

All patients had medically refractory epilepsy as daily disabling seizures. The mean age of patients was 8.5 years (14 months-24 years) [Table 1]. Patients were evaluated in detail with clinical history and examination, prolonged video-electroencephalogram (VEEG), detailed neuropsychological testing, MRI brain, DTI, PET scan (n = 6), fMRI (n = 4), WADA test (n = 1) and invasive recording (n = 1). Of seven patients four had left sided pathology and three had right sided pathology. All patients except one underwent pure disconnection and one underwent partial resection. Conclusion: 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. In our series, all seven patient had good seizure outcome and none had functional disabilities.

Key Words

Posterior quadrant disconnection, posterior quadrant dysplasia, posterior quadrant epilepsy, refractory epilepsy

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Table 1: Clinical details, investigation findings and outcomes of the current series

| Age (years) / handedness | Clinical features and findings | Seizures Age at onset | MRI | VEEG-interictal | VEEG-ictal | Functional MRI | PET hypometabolism | Outcome (Engel’s Grade) (follow-up) |
|-------------------------|--------------------------------|-----------------------|-----|----------------|-----------|----------------|-------------------|-----------------------------------|
| 24/M/L                  | Perinatal insult, Divergent squint, mild R-hemiparesis, reduced dexterity R-hand and hemiataxia R-inferior temporal quadrant anopia | Multiple daily extratemporal seizures without aura 19 years | L-P-O gliosis | Multifocal and bihemispheric Non-specific slowing L posterior temporal-central-parietal regions | Uncertain laterisation-surface EEG Invasive-L P-O onset with rapid F-spread | R lateralized language | None | Grade I (3 years) |
| 11/F/R                  | Meningitis Neonatal seizures R-hemifield visual loss | Daily CPS with visual L-P-O-T gliosis auras 1 year | L-PH and generalized SWD | CPS of L-PHR onset | R-lateralized language Vision-R-O | L-PHR | Grade I (2.5 years) Improvement in neuropsychology functioning |
| 4/F/R                   | Mild delayed development | Daily CPS with early nystagmus 2 years | R-P-Q dysplasia | R-PHR slowing, IEDs | R-PHR CPS | Vision bilateral | Intercital: R-PHR hypometabolism Ictal R-lateral P-O hypermetabolism | Grade I (2 years) Improved neuropsychological functioning |
| 3/F/L                   | Developmental delay L-facial and neck nevus Mild R-hemiparesis Ataxic gait | Daily drop attacks 18 months | Hemimegalencephaly maximal abnormality over L-PHR | L-H PHR slowing, electrical status over L-PHR | L-H drops of PHR onset | None | Hypometabolism L-H maximal over L-PHR | Grade I (2 years) Improved neuropsychological functioning |
| 11/M/R                  | Delayed milestones L-hemianopia Squint Decrease d L-hand dexterity | Daily CPS with aura as giddiness 1 month | R-P-O-Q dysplasia | Slowing with reduced amplitude in R-PHR IEDs: R-O; R-H; L-F; Bi-F. | CPS R-PHR onset | Motor area anatomically activated | Hypometabolism R-H; maximal over R-PHR | Grade I (10 months) Improved neuropsychological functioning |
| 14 months/M/- Twin      | LSCS - due to premature rupture of membrane Delayed development | CPS Daily | R-P-O-Q dysplasia | Slowing in R-PHR IEDs: R-O-T, R-H | CPS R-PHR onset | None | Ictal SPECT: R temporal hyper perfusion | Grade I (8 months) Improved neuropsychological functioning |
| 6/M/L                   | Normal except mild language delay R-hemianopia Reduced dexterity R-hand | CPS with visual aura Daily CPS 2 years | Left PT-O white grey differentiation with volume loss - dysplasia | Bilateral PHR slowing and bioccipital and generalized spikes | CPS left P-HR onset | None | Hypometabolism in left-P-T | Grade I (2 years) Improved neuropsychological functioning |

M = Male, F = Female, R = Right, L = Left, P = Parietal, O = Occipital, T = Temporal, Q = Quadrant, CPS = Complex partial seizures, PHR = Posterior head region, SWD = Spike wave discharge, SPECT = Single-photon emission computed tomography, IEDs = Interictal epileptiform discharges, VEEG = Video-electroencephalogram, MRI = Magnetic resonance imaging, PET = Positron emission tomography, LSCS = Lower segment cesarean section
In addition the splenial fibers originating in the ipsilateral hemisphere were also sectioned. The procedure is carried out in following stages:

**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 [Figure 1a]. 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 [Figure 1b].

**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 fibers 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 [Figure 1c]. The arteries and veins on the cortex are preserved as far as possible.

All patients underwent disconnection. In one patient, partial parietal-occipital resection was performed during posterior quadrant (PQ) disconnection (case 3). Four patients underwent the procedure on left side. None had any acute postoperative complications. One patient had a single postoperative seizure, but has remained seizure free at 2 years follow-up. All patients continue to be under regular follow-up (8-29 months; mean follow-up of 17.5 months).

Visual field deficits and squint likely due to visual defects was observed preoperatively in all the patients. Patients underwent PQ disconnection either due to the radiological (MRI) or electrophysiological abnormalities (interictal or ictal) involving the occipital-parietal and temporal regions. Although fresh visual field deficits following surgery were likely in all the patients none reported disabling visual symptoms after surgery.

In our series, perinatal insult (2/7) [Figure 2 and 3], posterior quadrant dysplasia (PQD) (4/7) [Figure 4, 6 and 7], and hemimegalencephaly (1/7) [Figure 5] were the etiology for refractory epilepsy. In patients with PQD, the dysplasia
sparing the motor cortex, allowing a motor sparing PQ disconnection to be performed. In the subgroup with PQD, complex partial seizures (CPS) (4/4) with prominent early ocular component (3/4) were observed. In the patient with hemimegalencephaly, electrophysiological and radiological abnormalities predominantly involved the posterior quadrant.

Figure 3: Eleven-year-old girl with perinatal insult and daily refractory seizures manifesting as visual aura progressing to tonic asymmetric contractions (right more than left) with falls and left occipitoposterior temporoparietal gliosis on MRI [Case 2-Table 1]. (a and b) Interictal EEG showing generalized epileptiform discharges with attenuation over left posterior head regions. (c) VEEG captured left hemispheric complex partial seizures with generalized attenuation at seizure onset followed by 3-4 Hz generalized spike and waves. Postictal EEG showed diffuse slowing with increased attenuation over the left posterior head region. She was uncooperative for visual field assessment. (d) Functional MRI (f-MRI) for visual fields (checker box-D1 and cartoons-D2) showed BOLD activation only in the right occipital area. Language f-MRI (D3-sentence completion and D4-verb generation) showed language lateralized to the right hemisphere. Motor finger tapping BOLD activation matched anatomical hand area (D5). She underwent a left posterior quadrant disconnection to become seizure free (e-postoperative CT). Postoperative EEG showing normal sleep patterns without epileptiform discharges (f).

Figure 4: A 4-year-old girl with medically refractory daily seizures of 2 years duration [Case 3-see Table 1]. MRI showed right posterior quadrant dysplasia (a). VEEG captured right posterior head region seizures (b). Ictal PET showed increased metabolism in the right lateral parieto-occipital region (c). She underwent right posterior quadrant disconnection (d) to be seizure free at 2 year follow-up and is at school doing well.

Figure 5: 3-year-old child with left hemimegalencephaly (a-d) with anterior to posterior gradient (maximal affection over the posterior head region-radiological and EEG) and refractory daily drop attacks [Case 4-Table 1]. Interictal EEG showed focal status over the left posterior head regions (note left-right bipolar anterior to posterior montage) (e). Ictal data suggested extratemporal left hemispheric drops. She underwent left posterior quadrantectomy and is seizure free at 2 years follow-up with improvement in cognitive, language, and motor skills.

Figure 6: (a-c) Eleven year old boy [case 5-Table 1] with daily seizures. MRI brain (1.5 T) showing right posterior quadrant dysplasia (arrows) with suspicious white matter abnormality over the left frontal regions (arrow head). PET scan shows right hemispheric hypometabolism that is maximal over the right posterior head region (arrows), but normal on the left hemisphere (d, e and f). Interictal EEG (g) shows right posterior head region slowing as theta-delta activity (*), right posterior-temporal-occipital (PHR) (arrow), right frontal-temporal (short arrow) and less frequent left frontal epileptiform abnormalities. (f) VEEG recorded seizures from the right posterior head regions, as sequential right posterior temporal-occipital spikes (arrows) that evolves lateral over the right hemisphere (with prominent nystagmoid artifacts during evolution i-red arrows) to end over the right hemisphere (i-arrows) followed by postictal attenuation over the right hemisphere (k-*). DTI demonstrated abnormalities over right PHR only (l). The patient underwent right posterior quadrant disconnection to be seizure free (10 months). PHR = Posterior head region, DTI = Diffusion tensor imaging.
Seizures as extratemporal drops with early eye blinks were noted in this patient, who had effective seizure control with posterior quadrantectomy. Pediatric patient with perinatal insult had CPS with visual aura; while the adult had no aura and the seizures were classified as extratemporal drops with early ipsilateral head adversion.

Neurocognitive improvement was observed in all pediatric patients following surgery. All parents were gratified with the effects of surgery. The adult patient was employed effectively after control of his seizures.

Discussion

We here report a series of seven consecutive patients, who underwent PQ disconnection for refractory epilepsy. Of these one was adult and six were in the pediatric age group. All had good seizure outcome without postoperative complications. The etiologies responsible for refractory epilepsy included PQD, hemimegalencephaly, and perinatal insult-related ipsilateral porencephaly and gliosis. Hypomotor seizures without secondary generalization were predominantly observed in all the patients in the current series, as often observed in pediatric patients with early epilepsy.[4-6]

PQD is a sporadic cortical developmental malformation involving the temporal-occipital-parietal lobes and is often associated with early onset refractory epilepsy. PQD may be a representation of a more widespread cortical dysplasia or as hemi-hemimegalencephaly.[5] All patients in this group had early onset catastrophic refractory epilepsy with developmental delay, minor focal motor deficits, and visual field defects, which are often the clinical features of PQD.

PQD with electrophysiological abnormalities predominantly on the contralateral hemisphere can have negative impact on the surgical outcome. All except one patient had EEG abnormalities restricted to the side of surgery. In one patient with contralateral frontal epileptiform discharges, MRI suggested the possibility of a subtle contralateral frontal dysplasia. Interictal PET and DTI showed no abnormalities in this area. All seizures were recorded from the posterior cortex on the side of surgery. Postoperatively, this patient had grade I seizure outcome. In selected patients with minor MRI abnormalities on the contralateral hemisphere, interictal PET and DTI can help to characterize contralateral hemispheric abnormalities and project postoperative seizure outcomes. While minor MRI abnormalities of the contralateral hemisphere may not affect seizure outcomes, contralateral independent interictal EEG abnormalities may have bearing on both the seizure and the neuropsychological outcome after surgery.[7]

Posterior cortex gliosis secondary to perinatal hypoxic insult is a common cause of refractory epilepsy in developing countries. Children with birth asphyxia and perinatal brain injuries present with global developmental delay, impaired cognition, and seizures that are often medically refractory. Typically, these patients have bilateral posterior head region (PHR) epileptiform discharges on EEG, with ulegyria and bilateral gliotic changes on MRI. Surgical intervention is often not possible in view of bilateral involvement and tubular visual fields. Two patients (one adult) with refractory epilepsy secondary to perinatal insult and porencephalic cyst affecting one of the hemispheres were effectively treated with PQ disconnection. Although the adult patient had widespread interictal EEG abnormalities, all seizures were recorded from the PHR on the side of surgery and f-MRI had shown effective language lateralization to the healthy (right) side. In selected patients, with predominant unilateral involvement posterior cortex gliosis secondary to perinatal insult, good seizure outcome can be obtained by surgery.

Dominant generalized epileptiform discharges were seen in three children who underwent successful surgery. Generalized epileptiform abnormalities can occur in focal epilepsy affecting the pediatric age groups particularly if the lesion is congenital or acquired early. Many of these patients have favorable outcomes with focal resective epilepsy surgery and follow-up EEG demonstrate resolution of generalized epileptiform discharges.[8-9] Utilization of the normal physiological pathways, maladaptive neural plasticity and secondary epileptogenesis may explain presence of generalized epileptiform abnormalities in this subgroup with focal epilepsy.[6,10-12] Generalized EEG abnormalities are thus likely secondary phenomena that resolve after surgery.

PQ disconnection is demonstrated useful in refractory epilepsy even during the 1st year of life.[12] The youngest patient effectively treated in our series was 14 months. The technical details are very well-described in earlier literature, we used insular approach as described above in our patient series.[13]

In view of small very carefully selected patient series, with a relatively limited follow-up period it is difficult to compare with other published series in the literature. All patients were seizure free with reduced medications at the last postoperative follow-up. One patient who had early postoperative seizure was also seizure free at 2-year follow-up. The types and frequencies of surgical complications from our studies are
very similar to earlier series. Few studies have evaluated surgical outcomes in posterior cortex epilepsies (not selective for PQ disconnection). 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 etiology, 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.[13] Parietal resections fared worse outcome than occipital or parieto-occipital resections (52% seizure freedom vs 89 and 93%, respectively, at 5 years). Again patients with tumoral etiology 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**

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

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