Central quadrantotomy for intractable childhood epilepsy: operative technique and functional neuroanatomy

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Refractory subhemispheric epilepsy has been traditionally treated by resection. The last few decades have seen the emergence of disconnective techniques, for both hemispheric and subhemispheric disease. The aim of this study was to describe the technique for a disconnective surgery for large epileptogenic lesions involving the central (periorolanic cortices), parietal, and occipital lobes. This junctional cortex within the hemisphere (in contrast to anterior and posterior quadrantotomies) presents unique challenges when contemplating a complete disconnection of the region. The surgical technique is achieved through six distinct steps: fronto-central, inferior frontoparietal, lateral temporo-occipital, medial frontal, basal temporo-occipital, and posterior parasagittal callosal disconnections. The functional neuroanatomy of each step is described, along with cadaveric dissections. The authors describe this technique and include a case description of a young girl who presented with childhood-onset intractable epilepsy associated with cognitive stagnation. The postoperative seizure outcome in this patient remains excellent at 2 years’ follow-up, with gains in cognition and behavior. Excellent seizure outcomes can be achieved if the network encompassing the entire epileptogenic cortex is disconnected while ensuring preservation of fiber systems that link functionally eloquent uninvolved cortices adjacent to the central quadrant. https://thejns.org/doi/abs/10.3171/2022.11.PEDS22356

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tetailed neurological and neuropsychological evaluations should be performed to evaluate the cognitive age of the patient and the cognitive performances in different domains. Supplementary factors that should be considered are the educational level of the patient, along with his/her professional plans and hobbies. All these elements should be discussed with the patient and the family before detailing the surgical strategy, while considering the risk of a permanent impact of surgery on the patient’s quality of life. Secondary reorganization of the different cortical and subcortical networks could occur in patients with refractory epilepsy; thus, resting-state and task-based functional MRI (fMRI) represent useful tools to predict the location and modifications of eloquent cortices and associated pathways.

Central quadrantotomy is indicated in patients with an epileptogenic focus localized to the centro-parietal and occipital lobes. A hemispherotomy would be contraindicated in the presence of residual function in the temporal and anterior frontal lobes. In patients with central quadrantic epilepsy associated with a hemiparesis, the risk of motor worsening is considered unlikely. In cases of preserved motor function, this surgery can still be proposed if the fMRI shows localization of both motor areas to the contralateral hemisphere. Ideally, the fMRI should also confirm activation of the language areas to the contralateral hemisphere. In cases in which the language areas have not moved to the contralateral hemisphere, all attempts should be made to preserve the arcuate fasciculus (AF) and the ventral language stream during surgery. The inherent risk of an injury to these residual functions due to surgery should be evaluated in the context of the preexisting cognitive status of the patient and the seizure burden. In cases of residual function in the epileptogenic central quadrant, more restricted disconnections may need to be combined with or replaced by other contemporary alternative options, such as laser interstitial thermotherapy.7–10

In the patient described in this paper, Broca’s area was predominantly in the left hemisphere while Wernicke’s area and both motor areas were localized to the right hemisphere, enabling consideration of a surgery with efforts to preserve the AF.

**Case Illustration**

**History and Evaluation**

An 18-year-old left-handed girl presented with drug-resistant epilepsy from the age of 3.5 years after experiencing a prenatal stroke in the territory of the left middle cerebral artery. She was born at term without perinatal complications. At epilepsy onset, she had normal developmental and neurological examination results, except for a mild right-sided asymmetry of fine motor movements. Initially, she experienced focal motor type seizures with bilateral secondary generalization. She then developed seizures with impaired awareness and tonic/clonic features. The seizures had a right-sided predominance with up to 20 episodes in 24 hours and occurred increasingly during sleep. Phase I investigations at this stage showed a multifocal left hemispheric epilepsy with foci in the central lobe. Cerebral MRI confirmed the radiological sequelae of a left posterior middle cerebral artery stroke, with the majority of the involvement restricted to the centro-parietal and occipital lobes (Fig. 1). Interictal PET showed hypometabolism predominantly in these areas. Based on invasive EEG recordings that showed epileptiform activity primarily from the central lobe, a premotor cortectomy was performed at 7 years of age (in another institution) with subpial transection of the motor region. After this surgery, her neurological status worsened with a severe hemiparesis that subsequently improved with neurological re-education and rehabilitation. Following a short period of seizure reduction, she continued to experience numerous daily seizures with multiple seizure types that included focal sensorimotor (with frequent secondary generalization), absence, and psychomotor seizures. Her behavioral problems also worsened and cognitive plateauing occurred between the ages of 15 and 18 years. Neuropsychological analysis revealed an IQ of 46 (Wechsler Intelligence Scale for Children–Fourth Edition), corresponding to a cognitive age of 7 years.

Phase I evaluations (EEG and MRI) revealed abnormalities within the same zone as that observed prior to the first surgery. Functional MRI confirmed the presence of a bilateral dominance for language areas, with a dominance for Broca’s area in the left hemisphere and a dominance for Wernicke’s area in the right hemisphere. The fMRI for motor functions showed cortical representation for both hands in adjacent areas of the nondominant (uninvolved) hemisphere (Fig. 1). Phase II evaluations using stereo-EEG showed that the epileptogenic foci were localized to the centro-parietal and occipital lobes, with a predominance in the left precentral area and superior parietal lobe.

**Intra- and Postoperative Course**

The central quadrantotomy was performed guided by intraoperative electrocorticography (ECoG) and continuous scalp EEG monitoring. The disconnection was considered complete when EEG monitoring showed no further seizure propagation. An external ventricular drain was introduced into the lateral ventricle and kept closed during the 48 hours prior to its removal to enable early treatment of hydrocephalus and/or intraventricular hemorrhage. Postoperative MRI showed the anatomical isolation of the central quadrant (Fig. 2). The early postoperative clinical status was marked by an aggravation of the right hemiparesis and the appearance of conductive aphasia, which subsequently improved over time. Neurological examination 6 months after surgery showed that the hemiparesis had markedly improved and the aphasia had completely resolved. She was estimated to be in Engel class I (seizure free) at the latest follow-up, 2 years after surgery. In addition, her behavioral problems improved and she had a good quality of life, with independence in activities of daily living and progress in cognitive development and vocational training.

**Operative Technique**

**Craniotomy and Intraoperative Electrophysiology**

The patient’s head was fixed in a Mayfield three-point fixation system and turned to the contralateral side. A cra-
niotomy was performed to expose the perisylvian cortices with the most anterior aspect at the level of the anterior sylvian point. Somatosensory evoked potential responses were used to identify the central sulcus, further confirmed by monopolar stimulation of the precentral gyrus. Scalp EEG recordings were continuously monitored during surgery from ipsilateral anterior frontal and occipital electrodes, in addition to contralateral multilobar electrodes. In addition, ECoG was performed over the exposed cortices prior to and after completion of disconnection.

Disconnection Procedure and Functional Neuroanatomy

Fronto-Central Disconnection

The disconnection begins at the lateral convexity of the hemisphere and follows in a lateromedial direction (Figs. 3 and 4, Video 1).

VIDEO 1. Clip showing the principal surgical steps when performing a central quadrantotomy. ant asc ramus = anterior ascending ramus of the lateral sulcus; CP = choroid plexus. © Giulia Cossu, published with permission. Click here to view.

The fronto-central disconnection follows a trajectory perpendicular to the cortical surface to reach the roof of the middle third of the body of the lateral ventricle, running deep and parallel to the precentral sulcus and corticospinal fibers. The precentral sulcus is identified on the brain surface, and the incision starts just anterior to it in a perpendicular direction to the surface, in the direction of the lateral ventricle. When reaching the subcentral region, we attempt to spare the pars opercularis and keep the precentral junction area (ventral premotor area) intact. The aim of this maneuver is to preserve the language areas in the dominant hemisphere.

At this step, and on the way to the supero-lateral ependyma, the superior longitudinal fasciculus (SLF II and III) is cut, thus disconnecting two components of the associative tracts between the frontal and parietal lobes (Fig. 5A–C).

Inferior Frontoparietal Disconnection

The inferior frontoparietal disconnection starts just above the subcentral gyrus and runs posteriorly parallel and superior to the sylvian fissure. When reaching the supramarginal gyrus, the disconnection is moved inferiorly in a vertical direction, posterior to the posterior sylvian point. In the depth, it is directed toward the posterior third of the lateral ventricle anteriorly and atrium posteriorly, through an axial (suprasylvian) and coronal (supramarginal) oblique trajectory to spare the AF before entering the lateral wall of the ventricle through the perisylvian cortex. This cut disconnects the central lobe from the basal ganglia, brainstem, spinal cord, and thalamus, as well as the inferior parietal lobe from the temporal and superior parietal lobes. In this step, the projection fibers, namely the corticospinal tract and thalamocortical fibers, as well as...
the retrolentiform portion of the internal capsule, are disconnected. Deepening the incision in the direction of the posterior part of the lateral ventricle body is performed in a superiorly and posteriorly 45° directed oblique trajectory, preserving the AF while interrupting the SLF III and middle longitudinal fasciculus (MLF; Fig. 5C–F).

Lateral Temporo-Occipital Disconnection

The disconnection continues inferiorly in the coronal plane and slightly curves posteriorly to proceed toward the preoccipital notch. It is directed toward the lateral wall of the atrium, following a perpendicular trajectory to the cortex through a plane between the posterior aspect of the middle temporal gyrus and the preoccipital notch. It disconnects the occipital lobe from the temporal lobe while interrupting the vertical component of the SLF, the posterior extension of the inferior fronto-occipital fasciculus (IFOF), the posterior limb of the anterior commissure (AC), the sublentiform portion of the internal capsule (including the optic radiations), and the tapetum, thereby disconnecting the basal and lateral aspects of the occipital lobe from its homologous cortex (Fig. 5D–F).

Medial Frontal Disconnection

The medial frontal disconnection begins at the superior limit of the fronto-central disconnection and deepens to reach the interhemispheric fissure, where the falx will be visualized through the pia, and the supracallosal sulcus is reached. It disconnects the central and superior parietal lobes, as well as the posterior cingulum, from the frontal lobe and anterior cingulum by interrupting SLF I and the cingulate bundle, respectively (Fig. 6).

Basal Temporo-Occipital Disconnection

The basal temporo-occipital disconnection is performed as a continuum of the lateral temporoparietal window, in which an inferior oblique direction is followed to reach the pia of the basal temporo-occipital junction and the tentorium. The most mesial part of this disconnection reaches a basal point where the falco-tentorial junction is noted. At this point, the parahippocampal gyrus is preserved anteriorly and the lingual gyrus is disconnected posteriorly. This incision through the fusiform gyrus interrupts the inferior longitudinal fasciculus (ILF; Fig. 6).

Posterior Parasagittal Callosotomy

The posterior parasagittal callosotomy represents the only pure intraventricular step of this procedure. It represents the last step of the central quadrantotomy, joining the medial frontal, fronto-central, inferior frontoparietal, and lateral and basal temporo-occipital (mediobasal temporo-occipital [MBTO]) disconnections. This callosotomy lies in a parasagittal plane and remains curvilinear due to the shape given by the telencephalic flexure where the anterior portion is more medial and the posterior part reaches a more lateral position. It disconnects the posterior callosal fibers and the cingulum from the parahippocampal gyrus while crossing the isthmus. From a microsurgical per-
FIG. 3. Identifying external surface landmarks such as the precentral, intraparietal, and Jensen intermediate sulci, and the distal ramus of the sylvian fissure, allows one to delineate the superior, middle, and inferior frontal, pre- and postcentral, superior parietal, supramarginal, and angular gyri, as well as the preoccipital notch. The surgery can be divided into six steps: fronto-central, inferior fronto-parietal, lateral temporo-occipital, medial frontal, basal temporo-occipital, and parasagittal posterior callosal disconnection. A: Lateral view of a left hemisphere anatomical specimen showing the central quadrantotomy cortical incision (dashed line). B: Lateral view of a left hemisphere anatomical specimen showing the medial frontal (MF; blue), fronto-central (FC; green), inferior fronto-parietal (IFP; red), lateral (LTO; yellow), and basal temporo-occipital (MBTO; orange) disconnection planes. C: Mediobasal view of a left hemisphere anatomical specimen showing the cortical incision of the central quadrantotomy (dashed line). D: Mediobasal view of a left hemisphere anatomical specimen showing the MF, MBTO, and posterior parasagittal callosal (PPC; purple) disconnection planes. © Giulia Cossu, published with permission.

FIG. 4. Summary of the six distinct surgical steps and their correlation with the disconnected and spared fiber systems at each stage. Vertical c. SLF = vertical component of the SLF.
FIG. 5. Some important anatomical structures are highlighted for the white matter dissection technique on a left hemisphere. The dashed lines show the incisions used for the central quadrantotomy. The view from the lateral aspect shows the fronto-central (FC), inferior frontoparietal (FP), and lateral temporo-occipital (LTO) steps of the procedure. A: The fronto-central disconnection runs lateromedially and superoinferiorly through the whole extension of the precentral sulcus, pars opercularis, and precentral junction (ventral premotor) area (asterisk). B: The main white matter fiber tracts and deep nuclei of the lateral aspect in the frontal and temporal lobes have been exposed, showing the SLF, corona radiata (cr), and IFOF, as well as the putamen (put) and head of the caudate nucleus (hcn). C: The next step in lateral dissection shows the extensions of the internal capsule, also known as the corona radiata. The main bulk of the SLF and AF have been left intact to understand their 3D relationship. D: The vertical component of the SLF has been peeled off, uncovering three association fiber tracts: the SLF II and III, and the AF. The SLF II and III connect the parietal and frontal lobes in a longitudinal direction from anterior to posterior. The AF is the only nonlongitudinally oriented tract into this area and is shown in the dissection wrapping around the posterior sylvian point, connecting the posterior aspect of the superior and middle temporal gyri with the central and frontal lobes. The parieto-temporal component of the AF has been cut deep to the superior temporal gyrus. Therefore, the parietal extension of the retrolenticular part of the corona radiata is exposed dorsally, as well as the IFOF ventrally. The IFOF is the lateral-most layer of the so-called sagittal stratum, which will be progressively uncovered in the next steps. E: The ventral supramarginal fibers around the posterior insular point have been removed and the retrolenticular part of the internal capsule (cr), together with the dorsal external capsule projection fibers, is exposed deep to an area in between the long insular gyrus and the AF. The inferior peri-insular sulcus has been uncovered and the temporopontine projection fibers (sublenticular IC) are partially exposed (cr). SLF II and III, as well as the parieto-temporal component of the AF, have been cut deep to the postcentral sulcus. Some connections of the SLF II to the superior parietal lobe remain intact to appreciate its posterior extensions. The dissection has been moved ventrally toward the limen insulae. This maneuver allows identification of the sublenticular bundle of the AC (aco) after cutting the dorsal component of the IFOF and opening Gratiolet’s canal just ventral to the putamen (put). Therefore, the posterior limb of the AC is exposed running to the temporo-occipital neocortex into the sagittal stratum thickness. The AC bundle represents the intermediate component of the sagittal stratum, also intermingling with the most superficial IFOF, and deeper optic radiations (sublenticular part of the cr). F: The AC has been cut just below the putamen (asterisk) and carefully peeled away posteriorly to uncover the deepest component of the sagittal stratum, i.e., Meyer’s loop (M’lo) and optic radiations (or). The most anterior aspect of the temporal horn has been opened, showing the head of the hippocampus and the uncal recess separating the hippocampus and the amygdala. The retroentoriform part of the corona radiata has been carefully removed at the level of the atrium lateral wall. A very thin layer of fibers covering the atrial ependyma is seen running downward and forward; this is the lateral temporo-occipital component of the splenium of the corpus callosum (cc), known as the tapetum (Tap). aco = anterior commissure; ang = angular gyrus; cr = corona radiata; cs = central sulcus; ins = insula; mtg = middle temporal gyrus; ol = occipital lobe; pcc = precentral gyrus; ptg = postcentral gyrus; pts = postcentral sulcus; put = putamen; spl = superior parietal lobule. © Giulia Cossu, published with permission.
spective, this incision is placed in the medial wall of the lateral ventricle and extends between two reference points, namely point P (precommissural point, Pablo’s point) and point R (retrohippocampal), thereby severing the only remaining attachments of the central quadrant by cutting the splenium projections (forceps major), the fibers connecting the posterior cingulum to the precuneus, as well as the mediobasal “u” fibers in this area (Fig. 6).

Discussion

Epilepsy is a network disease in which the afferent and
afferent fiber systems linking the epileptogenic foci are responsible for the secondary spread and clinical manifestations of the disease. Multilobar epilepsy, which accounts for 12%–22% of the surgical series in the pediatric neurosurgery literature, is defined as an epileptiform activity involving two or more lobes of the brain. Common etiologies include perinatal insults, head trauma, and type II cortical dysplasia. Refractory epilepsy remains a challenging disease to treat; in childhood, it may be severe and associated with brain development retardation. Patients with refractory multilobar epilepsy respond well to early surgical procedures such as resection, or alternatively to tailored subhemispheric disconnective procedures, based on the localization and extensions of the epileptogenic focus. The seizure freedom rate reported in the literature after these procedures varies widely between 18% and 60%, with a poorer outcome for multilobar surgery when compared with lobar resections. Studies investigating the possible predictors of seizure recurrence are very scarce in the literature and show negative outcome predictors for dominant hemispheric lesions, especially for fronto-temporal or temporo-parietal resections, particularly those with incomplete resection evidenced on the postoperative MRI and/or residual epileptogenic activity on EEG. Patients with an extended occipital resection experience the best seizure-free rates at last follow-up, probably because of the higher likelihood of removing the epileptogenic zone. Furthermore, epilepsies arising from the more posterior areas have better localization due to the limited presence of spread patterns to other regions of the brain. Posterior cortex epilepsy surgery, involving the occipital and/or parietal lobes, accounts for 10% of focal resections in pediatric cohorts. A longer history of seizures before surgery was an independent predictor of seizure recurrence, possibly related to the epileptogenic networks in the immature brain, with secondary epileptogenesis. Completeness of resection of the epileptogenic zone is well correlated with seizure freedom. These procedures may result in improved quality of life, especially if performed early, because ongoing seizures might negatively affect neuroplasticity and neurodevelopmental outcomes in this population.

Disconnective surgery is based on the concept of isolating the primary epileptogenic cortex by interrupting the routes of spread within the epilepsy network. At the same time, it is imperative to ensure that the noninvolved subcortical white matter tracts that are subsuming important functions are preserved during the disconnection. Preoperative evaluations are then confirmed by intraoperative electrophysiological monitoring and correlations of surface anatomy with neuronavigation. Three-dimensional surface rendering and diffusion tensor imaging tractography are greatly important for adequate preoperative planning, choosing the proper cortical incision areas, and analyzing fiber direction. All these data can be incorporated into the neuronavigation system and facilitate the intraoperative orientation of such a large incision deep into the brain. When contemplating the surgical treatment of large epileptogenic lesions of the central (perirolandic), parietal, and occipital lobes, there are several peculiar and anatomical constraints that need to be considered. The neocortex of these areas lacks clear anatomical boundaries between the lobes. The complexity of this region is further increased within the deep white matter anatomy, and this is primarily related to the development of the telencephalic flexure, a major landmark that modifies the anatomy of the human brain with the more posterior areas becoming ventral and lateral. Associative fibers connecting the anterior areas with the posterior ones follow the flexure, thus becoming semicircular. In these areas, the projection, association, and commissural fibers intermingle, with some white matter tracts remaining curved and others longitudinal. A precise knowledge of the cortical and subcortical anatomy of this region is fundamental to successfully perform disconnective procedures of this region.

Conclusions

Central quadrantotomy represents a complete disconnection of the centro-parietal and occipital lobes from the frontal and temporal lobes. Clear anatomical boundaries between these lobes are lacking, while the subcortical anatomy is characterized by the curvilinear subcortical fiber systems. The anatomical study of the telencephalic flexure and the delineation of the relationships between the projection, association, and commissural fibers in this region are the key to achieve a complete disconnection of the central quadrant while preserving important cortices and tracts adjoining or traversing this intricate junctional area of the neocortex.

References


**Disclosures**

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

**Author Contributions**

Conception and design: Daniel, Cossu, Thomas, Marston, Messerer, González-López. Acquisition of data: Cossu, Aureli, Roulet-Perez, Thomas, Marston, Pralong. Analysis and interpretation of data: all authors. Drafting the article: Cossu, Thomas, Marston, Pralong, González-López. Critically revising the article: Daniel, Cossu, Aureli, Thomas, Messerer, González-López. Reviewed submitted version of manuscript: Daniel, Cossu, Aureli, Thomas, Messerer, González-López. Approved the final version of the manuscript on behalf of all authors: Daniel. Statistical analysis: Thomas. Administrative/technical/material support: Aureli, Thomas, Marston, Pralong.

**Supplemental Information**

**Videos**


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