Hemispherectomy

Article *in* Pan Arab Journal of Neurosurgery · October 2008

1 author:

Alaa Eldin Elsharkawy
University of traditional medicine, Yerev…

Hemispherectomy

Alaa Eldin Elsharkawy[1,2], Heinz Pannek[1], Falk Oppel[1]

Abstract

Introduction: Hemispherectomy became a more widely accepted surgical treatment for intractable epilepsy, secondary to severe unilateral hemisphere damage. Basic concept of the hemispherectomy changed from hemispheric resection in anatomical hemispherectomy to a less tissue removal with disconnection of the rest of the hemisphere in the functional hemispherectomy, up to minimal tissue resection with maximal disconnection in the hemispherotomy. This change away from extended resections to predominant disconnection methods lead to reduction of the complications and at the same time maintained the favourable seizure freedom rate.

Methods: In our current review, we review the hemispherectomy history and techniques in literature with details of our modified surgical technique of hemispherotomy. We also outline the indications, appropriate patient selection, and present our results in a large series of 74 patients who underwent hemispherotomy in our clinic between 1995 and 2006.

Results: In our clinical series, the 2-year follow-up shows that 72% of our patients (54 out of 74) had class I and II outcome according to Engel's classification system. No mortality occurred in the current series and postoperative complication was significantly lowered.

Conclusion: Hemispherotomy represents an efficacious, technically simple and safe surgical treatment for the management of patients with medically intractable seizures. *(p11-18)*

Key words: *Hemispherectomy, hemispherotomy, intractable epilepsy and seizures*

Introduction

History of hemispherectomy

Cerebral hemispherectomy is the surgical removal of one cerebral hemisphere leaving the basal ganglia undisturbed. Hemispherectomy was first introduced in 1928 by Dandy and L'Hermitte as a radical treatment for malignant glioma of one hemisphere.[3,26] Although the patients died from postoperative complications or disease progression, the degree of postoperative functional recovery was notable. Anatomical hemispherectomy for seizure was started in 1938 by the Canadian neurosurgeon Kenneth McKenzie, in a hemiplegic adult with medically resistant partial epilepsy.[21]

In 1950, Roland Krynauw, a South African neurosurgeon, reported the first major series of hemispherectomy, the excellent results gave this procedure worldwide popularity but with a failure to offer improvements in survival or quality of life compared with more conservative treatments.[20] Due to appearance of late-developing complications (hydrocephalus and superficial hemosiderosis) after a short benign course of hemispherectomy.[29] The classic anatomic hemispherectomy was largely abandoned in the 1970s and 1980s.[29]

Theodore Rasmussen, proposed in 1983, a modified "functional" procedure in which only the central portion of the cerebral hemisphere was removed to allow exposure and disconnection of the anterior frontal and posterior parieto-occipital poles and the corpus callosum.[26] The major advantage of Rasmussen techniques was reduction of the complication rate while the same rates of seizure freedom remained comparable to the anatomic procedure.[26]

In 1992, Villemure introduced the hemispherotomy technique; he reported the lateral hemispherotomy approach centred on the insula. The vertical approach was introduced in the same year by Delalande.[5,6] Schramm in 1995 introduced pre-insular hemispherotomy.[28,29] These innovations all involve initial tissue removal and disconnections of

[1]Neurosurgical Department
Bethel Epilepsy Centre
Bielefeld, Germany

[2]Neurosurgical Department
Cairo University
Cairo, Egypt

Correspondence:
Prof. Dr. Falk Oppel
Email: Falk.oppel@evkb.de
Dr. Alaa Eldin Elsharkawy
Email: alaa.elsharkawy@evkb.de

remaining structures.[5,6,28,29,32]

This change away from extended resections to predominant disconnection methods lead to the renewed wide application of the hemispherectomy in the treatment of the hemispheric epilepsy syndromes.[35]

Indication of hemispherectomy

Hemispherectomy is indicted in 3 different groups of lesion; the first group is the congenital lesions including the hemimegalencephaly, expanded cortical dysplasia, as well as the tuberoses sclerosis.[32] The second group is perinatal or early childhood acquired diseases, like classical indication of the infantile hemiplegia, hemiatrophy, porencephaly and other encephalic classic lesions. The third group is childhood acquired lesions; progressive hemiparesis with unilateral focal epilepsy, frequent epilepsia partialis continua, patients with Rasmussen encephalitis and after an inflammation attack.[11]

Presurgical evaluation and decision making

The Preoperative Protocol developed in our institute (Bethel Epilepsy Centre) to identify patients for surgery includes the patient's history and physical examination. Non-invasive video EEG monitoring is standard for all epilepsy surgery candidates to confirm the unilaterality of the seizure organ. The structural and functional neuroimaging include MRI and CT of the skull, PET for the metabolic status, and SPECT for the evaluation of the cerebral blood flow. To clarify the speech and memory lateralization the Wada test and F-MRI becomes accomplished in the healthy hemisphere. Neuropsychological evaluation and ophthalmological assessment completes the presurgical evaluation.

All findings are submitted in a multidisciplinary case conference for discussion. The participants of such a conference are neurologists, neuropaediatricians, neurosurgeons, neuroradiologists, psychologists, psychiatrists, and clinical neurophysiologists. The multidisciplinary case conference indicates if a patient is a candidate for a hemispherectomy or not. The decision of hemispherectomy is based on three outlined selection criterion. The first criterion is the pharmacoresistancy of the epilepsy syndrome.[24] The second criterion is a hemiparesis, contralateral to the epilepsy causing hemisphere. Finally, structural and functional changes must be found in the contralateral side to the hemiplegia.

Techniques of hemispherectomy

Hemispherectomy was conveyed in the literature as a general term and actually covers many procedures.[22]

Anatomical hemispherectomy

This surgery is based on complete cortical removal with or without ventricular opening. These procedures are also called classical hemispherectomy, hemidecortication, or hemicorticectomy.[22]

Dandy's technique depended on lateral ventricle opening. In his approach, the resection of the hemisphere started with ligation of the vessels, removal of the frontal lobe, and division of the corpus callosum.[3] Disconnection of the rest of the hemisphere is accomplished through an incision in the corona radiata to reach the depths of the temporal and occipital lobes. Resection of the temporal lobe mesial structures is considered standard today.[1]

Winston, et al described a technique keeping the lateral ventricle closed; the Sylvian fissure is opened and a vertical incision made across the parietal lobe, from the posterior part of the Sylvian fissure to the vertex. Subsequently, a plane of dissection is developed from the edges of the insula below the cortex and around the ventricles to the falx above the corpus callosum and the dura mater in the temporal fossa. Then, the cortical mantle is removed.[22,35]

Carson, et al described a technique by which the temporal, frontal, parietal, and occipital lobes are taken in sequence and the insular cortex aspirated to keep the ventricle closed.[1]

Functional hemispherectomy

This technique is based on partial anatomical resection and complete physiological disconnection of residual unresected brain.

It was described by Rasmussen; in his method, the temporal lobe and a central part of the frontal and parietal lobes, about the length of the corpus callosum, are removed. Then, the remaining frontal lobe and the occipital lobe are disconnected subpilally.[26]

Hemispherotomy techniques

This approach is based on maximal disconnection and minimal excision and can be viewed as radical hemispheric tractotomy. There are two different approaches.

Vertical approach: This approach was described by Delalande, et al; the aim of this approach is to reach the lateral ventricle through a parietal parasagittal corticectomy. Through the ventricle the mesial structures of the temporal lobe are removed and the frontal and occipital lobes disconnected.[5,6]

A modification of lateral approach was introduced by Danielpour, et al. His aim was to reach the lateral ventricle through an interhemispheric approach and, then, complete disconnection of the hemisphere.[4]

Lateral approach: This approach depends mainly on

Sylvian fissure as a route to reach the lateral ventricle. Schramm, et al and Villemure and Mascott in 1995 introduced "peri-insular hemispherotomy".[28,32] This technique starts with subpial resection of the supra-Sylvian and infra-Sylvian parts from the frontal and temporal lobes. Then, the insula is exposed and an incision made along the circular sulcus, through the white matter to expose the inner surface of the lateral and temporal ventricles. By this window, callosotomy, disconnection of the frontal and occipital lobes, and removal of the temporal lobe mesial structures are performed.[22]

Hemispherical deafferentation

This approach begins with temporal lobe resection; the lateral ventricle is exposed by cortical incisions in the temporal horn. Finally, hemisphere disconnection is performed from the intra-ventricular space.

Transsylvian keyhole functional hemispherectomy

Schramm, et al modified their approach in this variation where the Sylvian fissure is opened and disconnections performed from the ventricular inner surface, which was exposed by means of incisions at the edges of the insula.[29]

Suprainsular window

In these techniques a window opened by incision of the pia matter 5-8 mm above the Sylvian fissure, along the inferior frontal gyrus through the space medial cerebral artery and its branches were coagulated, the mesial structures of the temporal lobe removed and the rest of the hemisphere disconnected.[2]

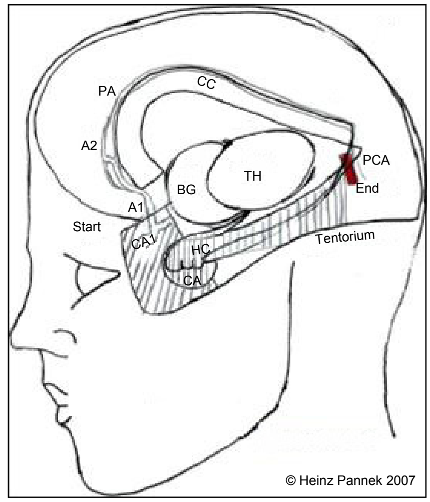
Our technique

In our institute we perform a modified technique of Delalande (1991); our own modified technique is a combined lateral and vertical hemispherectomy technique. The patient lies on his side with head straight on a horse-shoe. The skin incision for this pterional approach is a typical question mark incision, and over one burr-hole fronto-temporo-parietal osteoplastic craniotomy takes place with a diameter of approximately 6 cm. A free bone flap is turned to front-basal of the temporalis muscle. Before opening the dura, the operation microscope is introduced. The dural flap is cut in a satellite fashion with 2-3 additional relief cuts. The first step is to inspect the underlining pathology and landmark of the brain anatomy, the Sylvian fissure should be in the lower third of the craniotomy.

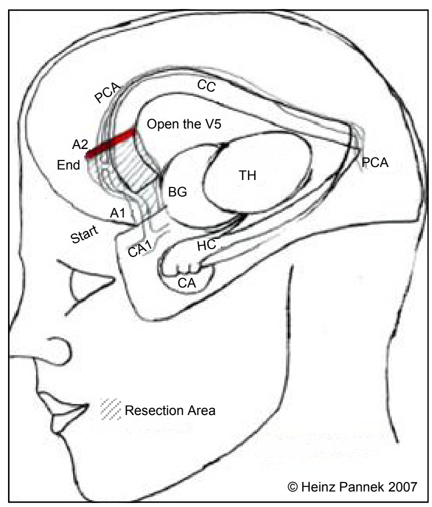
The operative procedures start with standard temporal resection, approximately 3-4 cm from the tip of the temporal lobe to the dorsal part. Subpial resection and suction with temperature-regulated bipolar-pincette of the superior temporal gyrus to the temporal base.

The dissection is then carried through the white matter at the uncus of parahippocampus gyrus and amygdala from rostral to basal direction. The hippocampus and parahippocampus are exposed and excised together with the fimbria of the hippocampus (Fig. 1).

Figure 1 - Schematic view of the first and second step



Step 1 - Standard temporal resection



Step 2 - ICA, A1 and A2 exploration

Starting from collateral eminence, a suction of ventricular parenchyma takes place up to tentorium. After completion of the temporal resection, it is preceded further to supra-Sylvian, just in front of the sphenoid wing; the fronto-orbital white matter is removed through a small incision with the sucker and with subpial suction technique from the anterior horn down to the frontobasal arachnoid in direction of internal carotid artery.

After reaching the ICA; both anterior cerebral arteries are exposed to identify the A1 artery on operative side and the A1 of the operative side is pursued anterior over the anterior communicating artery up to A2.

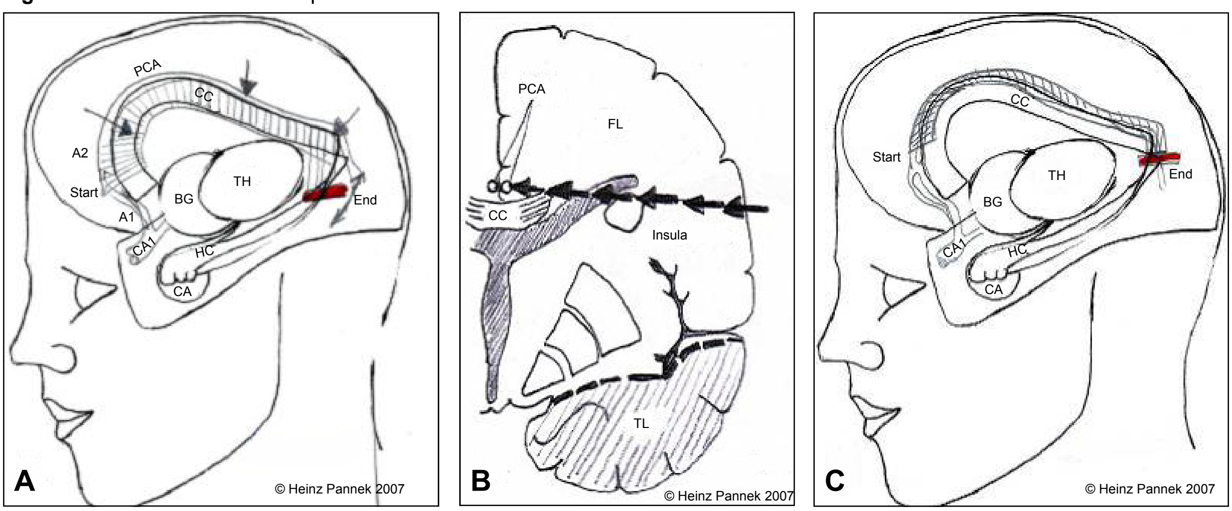
Using A2 as landmark: sucking away the rostrum of the corpus callosum, along the course of the pericallosal arteries around the corpus callosum (Fig. 2), disconnection of the frontal sections of the corpus callosum is then performed. The complete callosotomy is then started, including all of the splenium and the posterior third of the body of the corpus callosum, running along the inferior margin of the falx.

This is continued posteriorly until it joins the same line of dissection that has been prepared from the posterior aspect of the ventricular system; haemostasis is easily achieved by the placement of some oxidized cellulose gauze. All resected brain is sent for neuro-pathological examination, 2 hydro-static ventricle drainages placed and the dura is closed with the wound closed in layers.

Insular resection

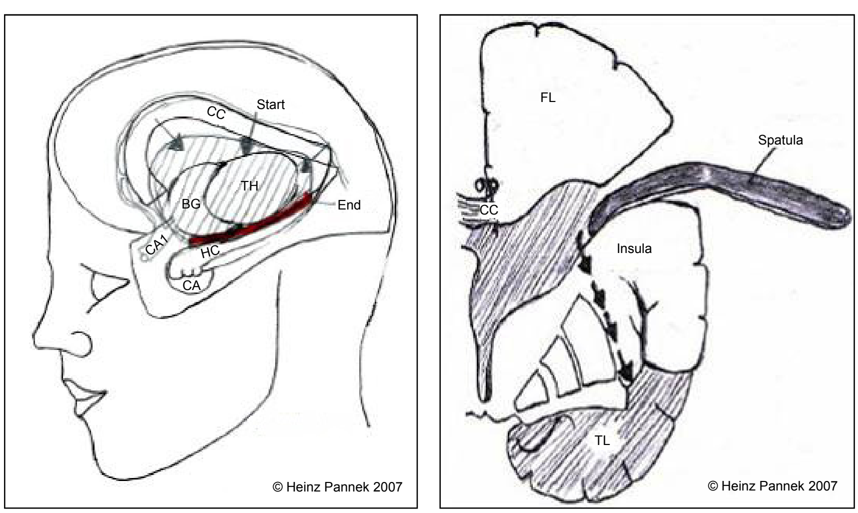
Insular resection is a standard in our institute, as it is the last step; patients with infarction usually had small or atrophied insula, patients with hemimegalencephaly have a large insular area, resection of insula start from above to separate the cortex of the insula away from the lateral ventricle (Fig. 3). Figures 4, 5, and 6 showing preoperative and postoperative

Figure 2 - Schematic view of step 3 and 4



Step 3 - (**a,b**) precallosal artery exploration. **Step 4 -** (**c**) fourth step corpus callosum disconnection

Figure 3 - Schematic view of the steps



Step 5 - Insular resection from above

MRI in three hemispherotomies; 6 month-old child with hemimegaloencephaly, 5 month-old child with cortical dysplasia and 2 year-old child with atrophy by infantile hemiplegia, respectively.

Results

Our series of 102 patient hemispherectomy procedures included 28 cases of functional hemispherectomy, 74 with our hemispherotomy technique (modified vertical technique). We retrospectively analysed the clinical profile and surgical outcome of the patients who had hemispherotomy.

There were 33 female and 41 male. The mean age at surgery was 6.4 ± 7 years range (0.25-34), mean epilepsy duration was 5.1 ± 6.2 years range (0.25-33.4). In our group there were 28 patients who had right-side hemispherotomy and 46 left-side hemispherotomy. Histopathological examination revealed that 14 (18.9%) patients had FCD, 4

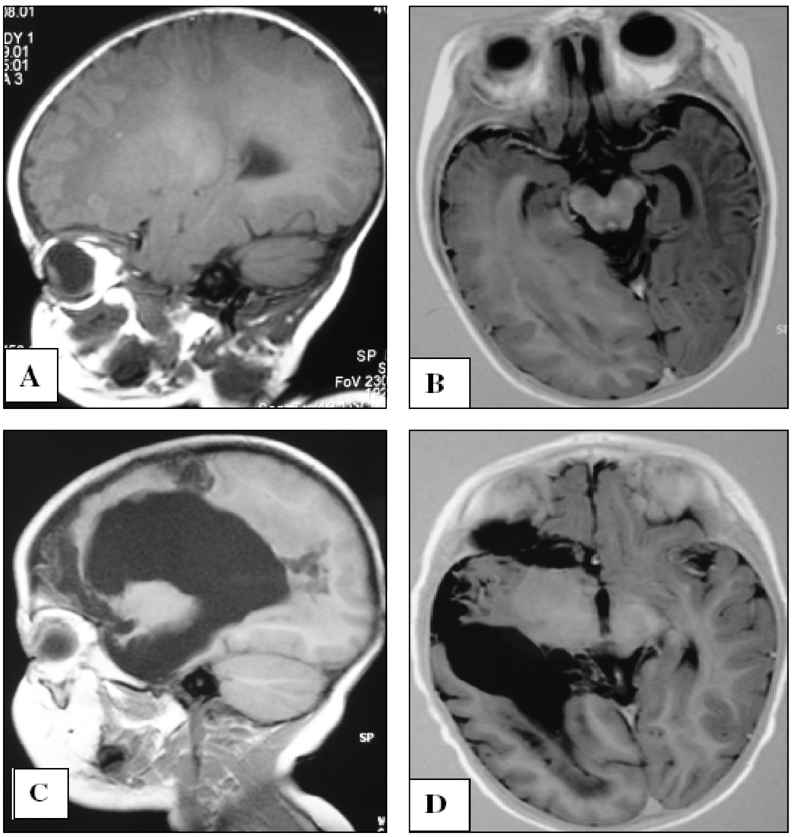


Figure 4 ← Schematic view: (**a,b**) Preoperative, (**c,d**) postoperative MRI for 6-month-old child with hemimegaloencephaly

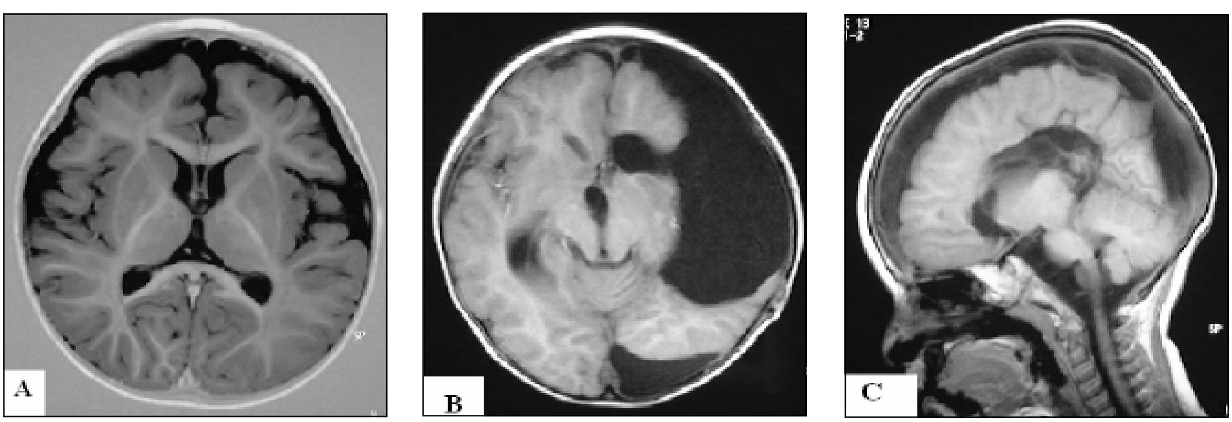


Figure 5 -Schematic view: (a) preoperative, (b,c) postoperative MRI for 5 month-old child with cortical dysplasia

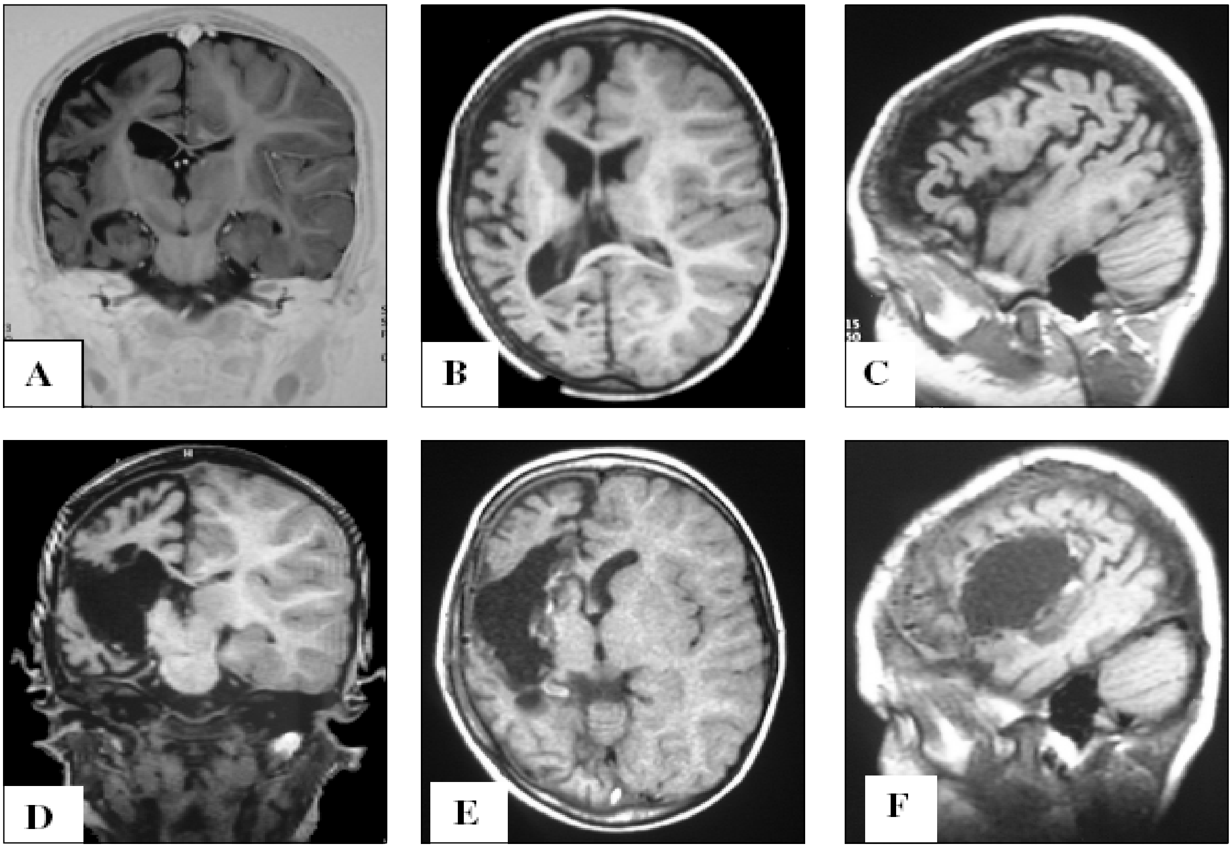


Figure 6 - Schematic view: (**a-c**) preoperative, (**d-f**) postoperative MRI for 2 year-old child with atrophy by infantile hemiplegia

(5.3%) had gliosis, 4 (5.3%) had atrophy, 14 (18.9%) had MCD, 8 (10.8%) had cerebrovasular accident, 7 (9.5%) had tuber sclerosis, 7 (9.5%) had Rasmussen syndrome, 8 (10.8%) hemimegaloencephaly, 5(6.8%) Sturge-Weber, 1 (1.4%) had neoplasm and 2 (2.7%) undetermined pathology. History of previous epilepsy surgery was recorded in 6 patients. In the group of hemispherotomy we had no mortality.

Seizure freedom rate was recorded in 72% of the patients (completely or nearly complete seizure free, Engel classification; class I and II) at 2 years follow-up. Seizure-free rates postoperative varied according to the pathology; 100% of patients with a Sturge-Weber syndrome, 76% of hemimegalencephaly, 72.7% in patients with cortical dysplasia, and 66.6% of acquired lesion, atrophy, porencephaly and Rasmussen-encephalitis, right-sided hemispherectomy had 75% seizure-freedom and left-sided 62% seizure freedom. In our series the following factors predicted the best postoperative outcome; higher presurgery developments and shorter epilepsy duration. Regarding complication in our series; 4% of the patients had postoperative wound infections, 8.8% showed incomplete disconnection in postoperative MRI. Shunt implantations amounted to 7% in our series and 9% of the patients required reoperation.

Discussion

Review of the published series in literature show that the seizure-free outcomes from other studies of hemispherectomy at 2 years follow-up vary from 54 to 88%.[6,19,18,24,26,30] Our results are in agreement with published results from other centres.[7,19,24,34] Our institute published the results of our hemispherectomy and hemispherotomy series; Holthausen (Oppel), et al 1997, 1999 and 2001, Tuxhorn and Pannek 2002.[13-16,31]

After successful hemispherotomy the decrease of seizure frequency and severity widens the scope of motor and social functioning, which overrides the effects of remaining motor impairments.[10] Moreover, Holthausen and colleagues from our institute found that patients who are ambulatory prior to hemispherectomy remain so thereafter, whether the pathology was acquired or not.[15,16]

As in previous series we found the early reference to surgery is a predicting factor for the best postoperative outcome with improvement of cognitive performance in most patients regardless of aetiology.[17,25]

The rate of the shunt implantations in our series appears acceptable and comparable with the VP shunt implantation rate in the other hemispherotomy series, in which 0%[(19)], over 8%[(33)], and up to 18%[(5)] are declared. Most cases of shunt implantation related to hemimegalencephaly patients.[8] In the literature, the reoperation rate varies between 5 and 19%.[2,15,23,27,30] In our series the rate of reoperation tends to be lower than the other series. There were no mortality in our hemispherotomy group, the mortality of the hemispherectomy fluctuates in published series between 0 and 3%.[5,7,14,29,34] In spite of the low complication rate hemispherectomy remains an intervention that has the highest morbidity and mortality rate in epilepsy surgery.[35]

Conclusion

Hemispherotomy is a successful neurosurgical treatment if the patient fulfils the criteria of intractable epilepsy with hemiplegia and diffuse functional and radiological hemispheric abnormality. The excellent outcome of hemispherotomy with acceptable rate of complication based on patient-selection criteria, the onward development in neuroimaging tools, the improvement of surgical techniques and surgeon's experience.

Short preoperative epilepsy duration is correlated to the favourable outcome, this finding provides direct support for promptly referring children with severe and medically resistant seizures for surgical evaluation; delay in referral could further worsen long-term neurodevelopmental status. Hemispherectomy, in the past regarded as a radical intervention of last resort, should become a standard of early intervention.

References

1. Carson BS, Javedan SP, Freeman JM, Vining EP, Zuckerberg AL, Lauer JA, et al: Hemispherectomy: A hemidecortication approach and review of 52 cases. J Neurosurg 1996, 84: 903-911
2. Comair YG: The transsylvian functional hemispherectomy: Patient selection and results. In: Lüders HO, Comair YG (eds): Epilepsy Surgery, 2nd Ed. Philadelphia, Lippincott Williams & Wilkins 2001, pp 699-704
3. Dandy WL: Removal of the right cerebral hemisphere for certain tumors with hemiplegia. JAMA 1928, 90: 823-825
4. Danielpour M, von Koch CS, Ojemann SG, Peacock WJ: Disconnective hemispherectomy. Pediatr Neurosurg 2001, 35: 169-172
5. Delalande O, Fohlen M, Jalin C, et al: From hemispherectomy to hemispherotomy. In: Lüders HO, Comair YG (eds): Epilepsy Surgery, 2nd Ed. Philadelphia, Lippincott Williams & Wilkins 2001, pp 741–746
6. Delalande O, Pinard JM, Basdevant C, Gauthe M, Plouin P, Dulac O: Hemispherotomy: A new procedure for central disconnection. Epilepsia 1992, 33:99-100
7. Devlin AM, Cross JH, Harkness W, Chong WK, Harding B, Vargha-Khadem F, Neville BG: Clinical outcomes of hemispherectomy for epilepsy in childhood and adolescence. Brain 2003, 126: 556-566
8. Di Rocco C, Iannelli A: Disconnective hemispherectomy. Pediatr Neurosurg 2002, 37:109
9. Döring S, Cross H, Boyd S, Harkness W, Neville B: The significance of bilateral EEG abnormalities before and after hemispherectomy in children with unilateral major hemisphere lesions. Epilepsy Res 1999, 34(1): 65-73
10. Empelen RV, Jennekens-Schinkel A, Buskens E, Helders PJM, Nieuwenhuizen OV: Functional consequences of hemispherectomy. Brain 2004, 127(9): 2071-2079
11. Feindel W: Theodore Brown Rasmussen (1910-2002): epilepsy surgeon, scientist, and teacher. J Neurosurg 2003, 98: 631-637
12. Hoffman HJ, Hendrick EB, Dennis M, Armstrong D: Hemispherectomy for Sturge-Weber syndrome. Childs Brain 1979, 5: 233-224
13. Holthausen H, May TW, Adams CTB, Andermann F, Comair Y, Delalande O, et al: Seizures post-hemispherectomy. In: Tuxhorn I, Holthausen H, Boenigk H (eds), Paediatric Epilepsy Syndromes and their Surgical Treatment. London, John Libbey 1997, pp 749-773
14. Holthausen H, Pieper T: Complications of hemispherectomy. In Lüders HO, Comair YG (eds), Epilepsy Surgery, 2nd Ed. Philadelphia, Lippincott Williams & Wilkins 2001, pp 879-883
15. Holthausen H, Strobl K, Pieper T, Teixeira VA, Oppel F: Prediction of motor function post hemispherectomy. In: Tuxhorn I, Holthausen H, Boenigk H (eds), Paediatric Epilepsy Syndromes and their Surgical Treatment. London, John Libbey 1997, pp 785-798
16. Holthausen H, Strobl K: Modes of reorganization of the sensorimotor system in children with infantile hemiplegia and after hemispherectomy. Adv Neurol 1999, 81: 201-220
17. Jonas R, Nguyen S, Hu B, Asarnow RF, LoPresti C, Curtiss S, de Bode S, et al: Cerebral hemispherectomy: Hospital course, seizure, developmental, language, and motor outcomes. Neurol 2004, 62: 1712-1721
18. Kanev PM, Foley CM, Miles D: Ultrasound-tailored functional hemispherectomy for surgical control of seizures in children. J Neurosurg 1997, 86: 762-767
19. Kestle J, Connolly M, Cochrane D: Pediatric peri-insular hemispherotomy. Pediatr Neurosurg 2000, 32: 44-47
20. Krynauw RA: Infantile hemiplegia treated by removing one cerebral hemisphere. J Neurol Neurosurg Psychiatry 1950, 13: 243-267
21. McKenzie KG: The present status of a patient who had the right cerebral hemisphere removed. JAMA 1938, 111: 168-183
22. Nogueira De Almeida A, Marino R Jr, Aguiar PH, Teixeira MJ: Hemispherectomy: a schematic review of the current techniques. Neurosurg Rev 2006, 29(2): 97-102
23. Peacock WJ: Hemispherectomy for the treatment of intractable seizures in childhood. Neurosurg Clin N Am 1995, 6(3): 549-563
24. Peacock WJ, Wehby-Grant MC, Shields WD, et al: Hemispherectomy in children: a report of 58 cases. Childs Nerv Syst 1996, 12: 376-384
25. Pulsifer MB, Brandt J, Salorio CF, Vining EP, Carson BS, Freeman JM: The cognitive outcome of hemispherectomy in 71 children. Epilepsia 2004, 45: 243-254
26. Rasmussen T: Hemispherectomy for seizures revisited. Can J Neurol Sci 1983, 10: 71-80
27. Schramm J: Hemispherectomy techniques. Neurosurg Clin N Am 2002, 13: 113-134
28. Schramm J, Behrens E, Entzian W: Hemispherical deafferentation: An alternative to functional hemispherectomy. Neurosurg 1995, 33: 509-516
29. Schramm J, Kral T, Clusmann H: Transsylvian keyhole functional hemispherectomy. Neurosurg 2001, 49: 891-900

30. Shimizu H, Maehara T: Modification of peri-insular hemispherotomy and surgical results. Neurosurg 2000, 47: 367-373
31. Tuxhorn IE, Pannek HW: Epilepsy surgery in bilateral Sturge-Weber syndrome. Pediatr Neurol 2002, 26(5): 394-397
32. Villemure JG, Mascott CR: Peri-insular hemispherotomy: Surgical principles and anatomy. Neurosurg 1995, 37(5): 975-981
33. Villemure JG, Vernet O, Delalande O: Hemispheric disconnection: Callosotomy and hemispherotomy. Adv Tech Stand Neurosurg 2000, 26: 25-78
34. Vining EPG, Freeman JM, Pillas D, Uematsu S, Carson BS, Brandt J, et al: Why would you remove half a brain? The outcome of 58 children after hemispherectomy - The Johns Hopkins experience: 1968 to 1996. Pediatrics 1997, 100(2): 163-171
35. Winston KR, Welch K, Adler JR, Erba G: Cerebral hemicorticectomy for epilepsy. J Neurosurg 1992, 77: 889-895