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Case Report

Functional Hemispherotomy in Medically Refractory Childhood Epilepsy Secondary to Perinatal Asphyxia-An Institutional Experience

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- Drug resistant epilepsy
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Abstract

Introduction: Drug resistant epilepsy is difficult to treat. Surgical management was first introduced by Dandy in 1928 when he performed the first anatomical hemispherectomy for infantile seizures. The treatment was very effective in the management of seizures but was associated with significant complications such as hydrocephalus, hemiplegia, aphasia and cortical hemosiderosis which develops later and is the cause of significant disability. The procedure now has been gradually replaced by functional Hemispherotomy where the epileptiform foci in the affected hemisphere are disconnected from the rest of the brain thereby containing the electrical discharges and preventing their translation into physical symptoms. This clinical remission from disabling epilepsy is highly successful with minimal complications in the short and long term to the patient.

Case series: We present our experience in a case series of 3 patients who underwent this procedure at different ages for hypoxic insults suffered in the immediate perinatal period. They were subjected to different combinations of antiepileptic drugs, but stayed away from surgery for different reasons. All underwent functional Hemispherotomy for their epilepsy. Post op they all had deficits which gradually improved to complete resolution after 3-6 months of physiotherapy.

Conclusion: For the right indications, Hemispherotomy is the correct surgical choice for disabling drug resistant seizures and offers good clinical improvement. Irrespective of the technique used, proper pre-operative assessment coupled with proper post-operative follow up and postoperative rehabilitative care are critical in ensuring complete return to functionality at the earliest.

INTRODUCTION

Functional Hemispherotomy as the name suggests implies a disconnection of a pathological hemisphere from the rest of the brain and its efferent fibres effecting a clinical remission of intractable epilepsy by containing the pathologic electrical activity inside the same hemisphere. [1] Such a concept was considered dangerous and too morbid to lead to any perceivable benefits. After the structural hemispherectomy where resection of the entire hemisphere was performed, complications were immense limiting the spread and usage of this procedure [1,2]. Disconnection as an alternative to excision proved much more acceptable as it reduced the need for disabling resections and its complications [1,2]. Today's disconnection Hemispherotomy is safe and considered an excellent surgical option for medically intractable epilepsy involving a diffuse focus or multiple foci localised to one Cerebral hemisphere [2]. The advent of Endoscopic transcallosal transventricular disconnection made this procedure even more acceptable by reducing the incision and craniotomy size making post op recovery easy and pain-free. We present our experience with a case series of 2 patients who underwent disconnection and performed well.

CASE REPORT 1

A 5 year old child presented to the clinic with refractory generalised seizures for 5 years. She had been on a full dosage of carbamazepine, levetiracetam and valproate for this period and had consulted various neurologist for her seizures. The child had a history of premature birth with an unmonitored antenatal period with perinatal meningitis needing ICU care for 3 weeks before recovery and discharge. On examination she was found to have a left hemiparesis of 2/5 power with left Homonymous hemianopia. Mental development was significantly retarded but speech was normal. AN MRI scan of the brain showed extensive polymicrogyria and gliosis pertaining to the old hypoxic and infectious insult (Figure 1a). The Video EEG recordings showed right frontocentral and temporal foci for the seizures. 2 seizures were recorded during a 24hr period with origins localised to the same focus. The lesions as shown extended almost across the entire hemisphere. She was evaluated and her relatives

were counselled for surgery. The lateral (Villemure) approach was selected due to surgeon comfort and proficiency. Motor mapping was done with on table Electrocorticography (ECoG) suprainsular dissection was carried out with an anterolateral and mesial temporal lobectomy, followed by insular and corpus callosal disconnection. Post op the child was extubated slowly in the ICU and covered with anticonvulsants. (Figure 1b) In the immediate post-operative period her deficits worsened into 0/5 power in the left side. Gradual physiotherapy led to improvement in power allowing her to walk unaided and use her left hand more and more (Figure 1c).

CASE REPORT 2

A 12 year of boy presented to the clinic with left sided hemiparesis and intractable epilepsy for the past 10 years. He was on carbamazepine, levetiracetam, phenytoin and valproate for variable periods in different combinations over 7 years. His neuropsychological assessment and rehab assessment indicated mild developmental regression. However he was ambulant and independently performing his household activities.

His MRI revealed extensive gliosis pertaining to perinatal asphyxia. (Figure 2a and b) A Functional MRI was performed to localise the motor strip as well as to ensure that the seizure origin localised to the affected hemisphere and lesion (Figure2c). After appropriate assessment and counselling, he was taken up for a disconnection procedure. A lateral approach was taken and a suprainsular dissection was done followed by an infrainsular disconnection. The gliotic tissue was dissected and excised as much as possible without disturbing the normal looking brain. Post operatively he developed a complete left sided hemiplegia

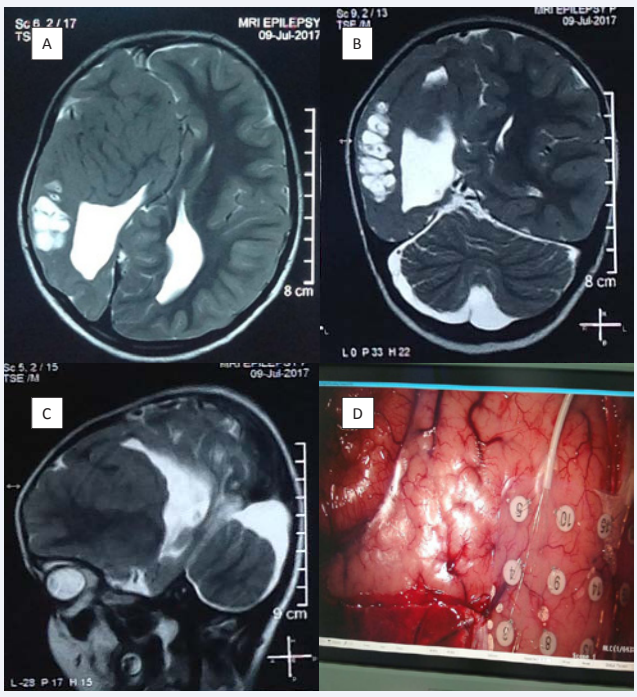

Figure 1 Preop Axial (A) Coronal (B) and Sagittal (C) MRI images of the child showing extensive gliosis and polymicrogyria. Intraoperative electrocorticography-ECoG (D).

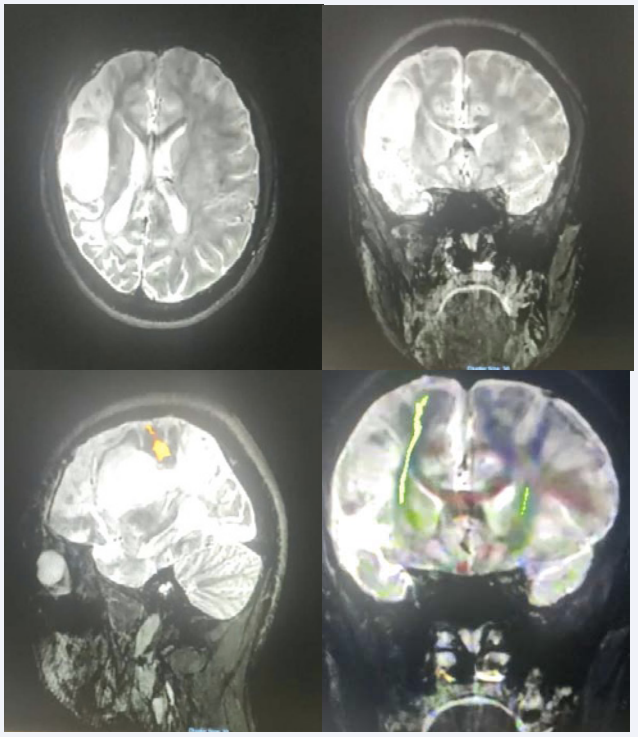

Figure 2 Pre op MRIs of the patient in (A) axial, (B) Coronal and Sagittal (C) views showing extensive hemispherical damage and gliosis. The Diffusion Tension Imaging (DTI) of the same patient shown in (D) shows the corticospinal tracts just deep to the gliotic region.

(0/5 power) but on physiotherapy he returned to preop power and independent mobility after 6months. (Figure 2d)

DISCUSSION

Functional disconnection as a surgery of choice for intractable epilepsy where diffuse seizure foci cannot be disconnected or excised individually[2-5]. The surgery employs advanced techniques used to disconnect the diseased hemisphere or part of the hemisphere from the rest of the functioning brain thereby limiting the clinical expression of the epileptiform foci of the hemisphere causing clinical remission of epilepsy [3]. Various technical embellishments have been made to the technique allowing more and more disconnection to be made with smaller approach corridors, the latest being the endoscopic disconnection Hemispherotomy. Nevertheless, the fundamentals of the procedure remain the same.

EVOLUTION OF THE PROCEDURE

Hemispherectomy was first introduced as a technique for intractable epilepsy where the focus is diffuse but confined to one hemisphere [1-4]. Dandy performed the first structural Hemispherotomy in 1928 for gliomas and 1938 for epilepsy and proved its efficacy [3]. Wiebe published a series in 2001 where he emphatically demonstrated the clinical remission of the disease post-surgery [3]. Surgical considerations and complications such as superficial cortical hemosiderosis were explained by Oppenheimer & Griffith in their landmark paper in 1966 [3, 4]. Rasmussen, Delalande, Villemure, Bucy, Schramm,

Cook and others first experimented with disconnection as the major component of the procedure as opposed to structural excision [3]. Delalande published his novel technique gleaned of the observation of butchery techniques of muscles to reduce the craniotomy yet hasten and improve the quality of the disconnection [4]. Bahulayen achieved international note when he demonstrated the feasibility of a purely endoscopic disconnection on cadavers [4].

Indications for functional Hemispherotomy:

The indications are few but specific. These include:

1. The presence of a diffuse focus or multiple foci of epilepsy occupying the greater part of and confined to one hemisphere [3].
2. Cortical malformations covering a large part of one hemisphere associated with epilepsy [3-5].
3. Presence of post-surgical or post traumatic gliosis across the hemisphere associated with medically intractable epilepsy [4].

Medically Intractable epilepsy (The rule of 2):

This involves the presence of physical or electrophysiological seizures in at least 2 episodes per year despite the administration of 2 or more antiepileptic's in full strength for 2 or more years and confirmed by 2 epileptologists after a full and thorough examination [3].

Pre-operative assessments: All phase 1 investigation such as EEG, Video EEGs, MRIs and even advanced imaging techniques such as PET, SPECT, MEG and Functional MRIs can be done to localise the focus. Once diagnosed and localised, various assessments peripheral to the diagnosis and localisation of the disease are performed [4, 5]. These include:

1) A visual assessment to document field losses prior to surgery. Due to the disconnection, there will be field losses varying from a superior lateral quadrantanopia (pie in the sky) to a full hemianopia.

2) Neuropsychiatric assessment of the effects of the epilepsy as well as a full IQ and investigations of differing types of mental retardation.

3) Physiotherapeutic assessment which will assume significance after surgery when a hemiplegia or hemiparesis will develop. This gradually improves with therapy.

Outine pre anaesthetic evaluation by a qualified and specialised neuroanaesthetist is also done to ensure a smooth and eventful procedure. Once all the assessment is complete, the family is introduced to the entire epilepsy team and a group counselling session is performed [6,7]. Once there is trust and all doubts and worries have been addressed an informed consent is taken for the procedure.

Functional hemispherical disconnection is divided into broadly 3 types: [7]

1. Periinsular disconnection
2. Frontal vertical disconnection
3. Endoscopic transcallosal transventricular disconnection

Peri-insular disconnection: (Figure 3a)

This was the traditional technique expounded by Wiesse initially and modified by Rasmussen and Villemure. [7-9] Cook further added by ligating the middle cerebral artery along with dissection and excision of the Insula which has now fallen into disuse. Today's lateral Hemispherotomy largely follows the technique of Villemure. This process involves a large fronto-parieto-temporal craniotomy centred around the sylvian fissure, following which 3 steps are employed. [8,9] The FIRST is to employ the suprainsular window disconnecting the frontal, parietal and occipital lobes from the deep nuclei of the hemisphere, followed by the SECOND step, where the infrainsular window is employed to disconnect the temporal lobe. This is often replaced by a anterolateral temporal lobectomy along with the removal of the mesial structures around the temporal horn of the lateral ventricle. FINALLY, the insula is excised leaving behind a completely disconnected hemisphere [8, 9].

Vertical Frontal disconnection: (Figure 3b)

This technique was popularised by Delalande, who used a small frontal incision to achieve complete hemispherical disconnection. [10] He proposed, the head to be in vertical position (neutral) and a small frontal craniotomy be made. The frontal horn of the lateral ventricle is dissected out and then the deep nuclei are identified. [10] The disconnection is then vertically done lateral to the deep nuclei until the choroid plexus of the temporal horn of the lateral ventricle is seen. Finally disconnection is done anterior to the frontal horn onto the floor of the anterior cranial fossa leading to total disconnection of the hemisphere. [10] This procedure is advantageous over the lateral disconnection as there is a smaller craniotomy and scalp flap dissection thereby making the post-operative recovery pleasant and quick. The disconnection achieved is as effective and complete as in the lateral procedure [10].

Endoscopic transcallosal transventricular disconnection:

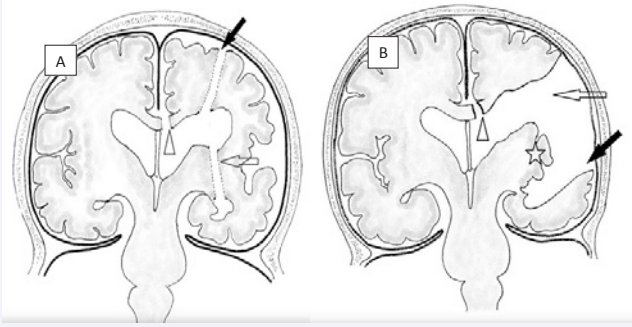

Figure 3 showing the techniques of hemispherical disconnection. In figure (A) we have the vertical disconnection technique of Delalande and in figure (B) the peri-insular dissection technique of Villemure. Pictures are courtesy of Bahuleyan B, Manjila S, Robinson S, Cohen AR. Minimally invasive endoscopic transventricular hemispherotomy for medically intractable epilepsy: a new approach and cadaveric demonstration. J Neurosurg Pediatr 2010;6:536-40.

This newly proposed technique involves a parasagittal craniotomy and a callosotomy into the ventricle using an endoscope [10-12]. The scope then follows the Delalande technique of ventricle disconnection into the temporal horn of the lateral ventricle. The technique uses a smaller incision and is helpful in children where hypothermia, infection and postoperative pain are major considerations in long and complex surgery of the brain [12].

Post-operative care: After surgery the patient is admitted into the neurosurgical ICU for monitoring. The anticonvulsants are continued as before surgery and fluid balance is maintained. Feeding might be started after 24 hours and once the patient is shifted to the ward to start his or her rehabilitation. Immediate concerns are seizures which maybe intractable and progress into status epilepticus. [12-14] Aggressive antiepileptic medication and anaesthesia must be used to reduce brain swelling including a generous use of steroids. Hydrocephalus maybe a concern but is addressed by regular clinical and radiological examination [13-15].

Long-term rehabilitative care is most important to the patient. Both physical and psychological care and support assist in a better and quicker recovery. [15] Epilepsy recovery is assessed on the Engel score where Engel 1, 2 and 3 indicate meaningful recovery while the Engel 4 and 5 imply no improvement or worsening of the disease. [16-18] Antiepileptic medication in time can be tapered and stopped or modified into monotherapy in a lower dosage making compliance better. [16-18]

CONCLUSION

Functional Hemispherotomy is considered an effective and useful technique to control diffuse foci of epilepsy localised to a single hemisphere. Correct assessment and a good team approach help in delivering the optimum results to the patient.

REFERENCES

1. Bahuleyan B, Manjila S, Robinson S, Cohen AR. Minimally invasive endoscopic transventricular hemispherotomy for medically intractable epilepsy: a new approach and cadaveric demonstration. J Neurosurg Pediatr 2010; 6: 536-540.
2. Basheer SN, Connolly MB, Lautzenhiser A, Sherman EM, Hendson G, Steinbok P. Hemispheric surgery in children with refractory epilepsy: seizure outcome, complications, and adaptive function. Epilepsia 2007;48: 133-140.
3. Carreño M, Wyllie E, Bingaman W, Kotagal P, Comair Y, Ruggieri P. Seizure outcome after functional hemispherectomy for malformations of cortical development. Neurology 2001;57: 331-333.
4. Shon W Cook , Snow T Nguyen, Bin Hu, Sue Yudovin, W Donald Shields, et al. Cerebral hemispherectomy in pediatric patients with epilepsy: comparison of three techniques by pathological substrate in 115 patients. J Neurosurg 2004;100(2 Suppl Pediatrics): 125-141.
5. Cross JH, Jayakar P, Nordli D, et al. Proposed criteria for referral and evaluation of children for epilepsy surgery: recommendations of the subcommission for pediatric epilepsy surgery. Epilepsia 2006;47: 952-959.
6. de Almeida AN, Marino R Jr, Marie S, Aguiar PH, Teixeira MJ. Factors of morbidity in hemispherectomies: surgical technique x pathology. Brain Dev 2006;28: 215-222.
7. Olivier Delalande ,Christine Bulteau, Georges Dellatolas, Martine Fohlen, Claude Jalin, Virginie Buret, et al. Vertical parasagittal hemispherotomy: surgical procedures and clinical long-term outcomes in a population of 83 children. Neurosurgery 2007;60:(2 Suppl 1): ONS19-32; discussion ONS32.
8. J Engel Jr, S Wiebe, J French, M Sperling, P Williamson, D Spencer,et al. Practice parameter: temporal lobe and localized neocortical resections for epilepsy: report of the Quality Standards Subcommittee of the American Academy of Neurology, in association with the American Epilepsy Society and the American Association of Neurological Surgeons. Neurology 2003;60: 538-547.
9. González-Martínez JA, Gupta A, Kotagal P, Elaine Wyllie, Hans O Lüders, William E Bingaman, et al. Hemispherectomy for catastrophic epilepsy in infants. Epilepsia 2005;46: 1518-1525.
10. Haruhiko Kishima , Satoru Oshino, Naoki Tani, Yomoyuki Maruo, Shayne Morris, Hui Ming Khoo, et al. Which is the most appropriate disconnection surgery for refractory epilepsy in childhood? Neurol Med Chir (Tokyo) 2013;53: 814-820.
11. Kwan A, Ng WH, Otsubo H, Ayako Ochi, O Carter Snead, Mandeep S Tamber. Hemispherectomy for the control of intractable epilepsy in childhood: comparison of 2 surgical techniques in a single institution. Neurosurgery 2010;67(2 Suppl Operative): 429-436.
12. Marras C, Granata T, Franzini A, Elena Freri, MarinaCasazza et al. Hemispherotomy and functional hemispherectomy: Indications and outcome. Epilepsy Res 2010; 89:104-112.
13. Morino M, Shimizu H, Ohata K, Tanaka K, Hara M. Anatomical analysis of different hemispherotomy procedures based on dissection of cadaveric brains. J Neurosurg 2002;97: 423-431.
14. Oppenheimer DR, Griffith HB. Persistent intracranial bleeding as a complication of hemispherectomy. J Neurol Neurosurg Psychiatry 1966;29: 229-240.
15. Pinto AL, Lohani S, Bergin A. Surgery for intractable epilepsy due to unilateral brain disease: a retrospective study comparing hemispherectomy techniques. Pediatr Neurol 2014;51: 336-343.
16. Schramm J, Kuczaty S, Sassen R, Elger CE, von Lehe M. Pediatric functional hemispherectomy: outcome in 92 patients. Acta Neurochir 2012;154: 2017-2028.
17. Villemure JG, Daniel RT. Peri-insular hemispherotomy in paediatric epilepsy. Childs Ner Syst 2006;22: 967-981.
18. Wiebe S, Blume WT, Girvin JP, Eliasziw M; Effectiveness and Efficiency of Surgery for Temporal Lobe Epilepsy Study Group. A randomized, controlled trial of surgery for temporal-lobe epilepsy. N Engl J Med 2001;345: 311-318.

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