

# CONTENTS

## Epilepsy surgery in children: Time is critical

J. Helen Cross, Alexis Arzimanoglou, Philippe Kahane, Hans Holthausen,  
Gary Mathern, William D. Gaillard, Prasanna Jayakar

| XIX

## Section I. PRESURGICAL EVALUATION IN CHILDREN

Editors: Prasanna Jayakar, William D. Gaillard

### Chapter 1: The role and limits of seizure semiology

J. Helen Cross

| 3

What can be determined from semiology in children?

| 3

Age dependence of semiology

| 4

### Chapter 2: The role and limits of surface EEG and source imaging

Prasanna Jayakar, Douglas Nordli, O. Carter Snead

| 7

Surface EEG

| 8

*General principles*

| 8

*Developmental substrates – Maturational issues*

| 9

*The epileptic generator*

| 10

*Technical issues*

| 11

*Practical “dos and don’ts” – Tips to avoid pitfalls*

| 11

Source imaging

| 15

*Background*

| 15

*Basic principles of source imaging*

| 15

*Localization of the epileptogenic zone in conjunction with other non-invasive neurophysiological and imaging modalities*

| 17

*Contribution to the decision making relevant to the indication for invasive monitoring*

| 19

*ESI/MSI: pros and cons*

| 20

*EEG-triggered fMRI*

| 21

### Chapter 3: The role and limits of structural and functional neuroimaging

William D. Gaillard, Chima Oluigbo

| 25

Structural imaging in presurgical evaluation

| 26

Functional imaging to identify the epileptogenic zone

| 29

Mapping eloquent cortex

| 32

### Chapter 4: The role and limitations of cognitive evaluation

Mary Lou Smith, Madison Berl

| 39

Purposes of the presurgical neuropsychological evaluation

| 40

Domains included in the neuropsychological evaluation

and guidelines for determining assessment tools within those domains

| 40

*School-age children*

| 41

*Considerations for young (preschool) and children with intellectual disability*

| 45

Intracarotid anaesthetic procedures in children

| 45

Challenges for and limitations of the presurgical neuropsychological evaluation in children

| 48

Future directions

| 48

### Chapter 5: The role and limits of behavioral and psychiatric evaluation

Jay Salpekar, David Dunn

| 51

Overlap of epilepsy and psychiatric conditions

| 51

Neurologic pathways to psychiatric illness

| 52

Common psychiatric comorbidities

| 53

*Attention deficit hyperactivity disorder (ADHD)*

| 53

*Anxiety and depression*

| 53

*Comorbidity in drug-resistant epilepsy*

| 54

Surgery in neurology and psychiatry

| 54

Role of neurobehavioral evaluation

| 55

Elements of neurobehavioral evaluation

| 55

Risk and benefit analysis

| 56

Limits of neurobehavioral evaluation

| 57

Research directions for the future

| 57

<b>Chapter 6: Intracranial EEG recordings and electrical stimulation</b>	
Laura Tassi, Prasanna Jayakar, Tom Pieper, Philippe Kahane	61
General indications	62
iEEG methods: subdural/depth, SEEG, ECoG	63
<i>Subdural electrodes and combination of subdural/depth electrodes</i>	63
<i>Stereo-electro-encephalography (SEEG)</i>	65
<i>Intra-operative electro-corticography (ECoG)</i>	68
Defining the epileptogenic zone (EZ): spontaneous and provoked	69
<i>Ictal onset zone</i>	70
<i>Irritative, continuous epileptiform discharges, and functional deficit zones</i>	72
Defining critical cortex: intra-operative and extra-operative	72
<i>Extra-operative electrical stimulation</i>	72
<i>Intra-operative electrical stimulation</i>	75
<b>Chapter 7: Why is the epilepsy case conference important?</b>	
Elaine Wyllie, Imad Najm	79
<b>Chapter 8: Socio-economic aspects and epilepsy surgery in children</b>	
Graham Fieggen, Jo Wilmshurst	83
What is required?	84
Developing (“resource poor”) countries	85
<i>Africa</i>	85
<i>South Asia</i>	87
<i>China</i>	88
<i>Middle East</i>	88
<i>Latin America</i>	88
Developed (“resource equipped”) countries	88
Specific paediatric issues	89

## Section II. SEMIOLOGY IN CHILDREN

Editors: Philippe Kahane, J. Helen Cross

<b>Chapter 9: The temporal lobe: Semiology</b>	
Andras Fogarasi	95
Ictal signs of temporal lobe seizures	95
<i>Behavioral change</i>	95
<i>Ictal emotional signs</i>	96
<i>Ictal motor signs</i>	96
<i>Autonomic symptoms</i>	96
Age dependency of different peri-ictal signs	96
Etiology and seizure semiology	99
Lateralizing signs in childhood temporal lobe seizures	99
Interobserver agreement on childhood seizure semiology	100
Further pitfalls of TLE seizure semiology	100
<b>Chapter 10: The frontal lobe: Semiology</b>	
Andras Fogarasi, Ingrid Tuxhorn, Philippe Kahane	103
Frontal lobe functional anatomy relevant to seizure semiology	103
Clinical patterns of frontal lobe ictal semiology	104
<i>Tonic seizures</i>	104
<i>Versive seizures</i>	104
<i>Hypermotor seizures</i>	106
<i>Motor stereotypies</i>	107
<i>Autonomic seizure</i>	107
Differential diagnosis and frontal lobe epilepsy syndromes	107
Do pediatric frontal lobe seizures and epilepsy differ from adults?	108
<b>Chapter 11: The posterior cortex: Semiology</b>	
Stefano Francione, Roberto Mai	111
Semiology of seizures originating from the parietal lobe	112
Semiology of seizures originating from the occipital lobe	113
Particular findings in (young) children	114
<i>Auras</i>	115
<i>Oculo-motor manifestations</i>	115
<i>Simple motor and complex motor behaviours</i>	116
<i>EEG features</i>	118

<b>Chapter 12: The insula: Semiology</b>	
Petia Dimova	121
Insular lobe semiology in children: is there any specificity?	124
<i>Auras</i>	126
<i>Vegetative signs</i>	126
<i>Motor manifestations</i>	126

### Section III. ETIOLOGY AND EPILEPSY SURGERY

**Editors: Hans Holthausen, Alexis Arzimanoglou, Prasanna Jayakar**

<b>Chapter 13: Focal (isolated) cortical dysplasia Type I</b>	
Hans Holthausen, Pavel Kršek, Ingmar Blümcke	133
Histological classification of FCD Type I – work in progress	133
Imaging in isolated FCD Type I	137
<i>MRI in isolated FCD Type I</i>	137
<i>PET and SPECT in isolated FCD Type I</i>	139
Seizures in FCD Type I	140
EEG in FCD Type I	140
Presurgical work-up and epilepsy surgery in FCD Type I	140
<b>Chapter 14: Focal cortical dysplasia Type II</b>	
Laura Tassi, Ingmar Blümcke, Deepak Gill	145
Histopathological and MR aspects of FCD type II	146
<i>Histopathological aspects</i>	146
<i>Neuroradiological methodology and diagnosis</i>	148
Clinical and neurophysiological aspects	149
<i>Clinical data</i>	149
<i>EEG and video-EEG features</i>	149
<i>Ancillary techniques</i>	150
Invasive techniques and surgical outcome	150
<i>Invasive monitoring: when and why</i>	150
<i>Surgery and outcome</i>	153
<b>Chapter 15: Other malformations of cortical development</b>	
Carmen Barba, Frank Ritter, Renzo Guerrini, Angus A. Wilfong	157
Heterotopias	158
<i>Subependymal (periventricular) nodular heterotopias</i>	158
<i>Subcortical nodular heterotopias</i>	159
<i>Subcortical band heterotopias</i>	161
Polymicrogyria	162
<b>Chapter 16: Tuberous sclerosis</b>	
Michael Duchowny, A. Simon Harvey, Howard Weiner	167
Clinical presentation	167
Surgical referral and pre-operative assessment	168
Intracranial EEG recordings	170
Surgical approaches	171
Outcome	173
<b>Chapter 17: Hemimegalencephaly and diffuse hemispheric malformations of cortical development</b>	
Christine Bulteau-Peyrie, Taisuke Otsuki, Olivier Delalande	175
Specific epilepsy surgery indications	175
Recommended presurgical evaluation	177
<i>Clinical considerations</i>	177
<i>Brain MRI</i>	177
<i>Interictal and ictal video EEG</i>	178
Available results	178
<i>Seizures</i>	178
<i>Contra-lateral hemiplegia and homonymous lateral hemianopia</i>	180
Suggested follow-up	181
<b>Chapter 18: Benign tumors (WHO grades I and II)</b>	
Hans Holthausen, Bertil Rydenhag, Nandan Yardi, Ingmar Blümcke	185
Neuro-pathological classification of brain tumors WHO grade I and II frequently associated with epilepsy (so-called LEATs)	186

MRI in patients with epilepsy in association with brain tumors	187
Incidences, prevalences, frequencies in pediatric brain tumors	190
Brain tumors and epileptogenicity	191
Medical therapy/AEDs in children with epilepsy in association with brain tumors	191
Surgery in children with seizures associated with benign tumors	192
<i>Tumor-surgery vs. epilepsy surgery and lesionectomy vs. tailored resection</i>	192
Radiotherapy and chemotherapy in patients with epilepsy and benign supratentorial brain tumors (WHO grade I and II)	199
Surgery in patients with epilepsy in association with brain tumors WHO grade II and higher	200
<b>Chapter 19: Vascular causes and perinatal hypoxic-ischemic events</b>	
Eeva-Liisa Metsähonkala, Hans Holthausen, Yu-Tze Ng, Eijja Gaily	207
Prenatal and perinatal lesions	208
<i>Arterial ischemic strokes and porencephalic cysts in children</i>	208
<i>Venous stroke and cerebral sinovenous thrombosis</i>	209
<i>Intracranial hemorrhage and hemorrhagic stroke in term infants</i>	209
<i>Watershed lesions and ulegyria due to hypoxic ischemic events</i>	210
<i>Vascular lesions in preterm children</i>	210
<i>Seizure types and epilepsy syndromes in pre/perinatal vascular lesions</i>	210
<i>Presurgical evaluation in pre/perinatal vascular lesions</i>	211
<i>Surgery in pre- and perinatal vascular lesions and lesions caused by hypoxic ischemic events</i>	215
<i>Postoperative outcome</i>	217
Epilepsy surgery in vascular lesions occurring in childhood	220
Cavernomas	221
Hematological testing for neurosurgery	223
<b>Chapter 20: Sturge-Weber syndrome</b>	
Alexis Arzimanoglou, Eric Kossoff	229
Clinical presentation	229
Diagnostic workup	230
Medical and surgical treatment	231
<b>Chapter 21: Hypothalamic hamartoma</b>	
John F. Kerrigan, Philippe Kahane, Martine Fohlen, Alexis Arzimanoglou	239
History	239
Epidemiology	240
Genetics	240
Anatomical aspects	241
Pathology and pathophysiology	241
Clinical-EEG features and natural history	243
<i>Gelastic and dactylic seizures</i>	244
<i>Other seizure types</i>	244
<i>Cognition and behaviour</i>	245
<i>Psychiatric symptoms</i>	246
HH and epilepsy: which part of the brain is seizing?	247
<i>Intrinsic epileptogenesis</i>	247
<i>Secondary epileptogenesis</i>	248
<i>The running-down phenomenon</i>	250
Presurgical evaluation	251
<i>Magnetic resonance (MR) imaging</i>	251
<i>Electroencephalography and video-EEG seizure monitoring</i>	251
<i>Neuropsychological or neurodevelopmental testing</i>	251
<i>Electrocorticography and invasive EEG monitoring</i>	251
Treatment	252
<b>Chapter 22: Cerebello-pontine hamartoma</b>	
A. Simon Harvey, Alexis Arzimanoglou	261
Clinical presentation	261
Pathological and functional considerations	262
Surgical treatment and outcomes	264
<b>Chapter 23: Epilepsies following cerebral infections</b>	
Hans Holthausen, Georgia Ramantani	267
Epilepsy post viral encephalitis	270
Epilepsy post-herpes simplex virus encephalitis	270
Epilepsies post-bacterial meningitis (EPBM)	275
Epilepsy in neurocysticercosis (NCC)	278
Epilepsy surgery in rare post-infection epilepsy	280

<b>Chapter 24: Rasmussen encephalitis</b>	
Adam L. Hartman, Cynthia F. Salorio	285
Historical perspective	285
Diagnosis	285
Pathology	288
Treatment	288
<i>Medical treatment</i>	288
<i>Surgery</i>	289
Outcomes	290
Timing of surgery	294
Post-surgical management	295
Directions for future research	295
<b>Chapter 25: Post-traumatic epilepsy</b>	
Brian J. Dlouhy, Matthew D. Smyth, David D. Limbrick, Jr.	299
Epidemiology	300
Pathophysiology	301
Treatment	301
<b>Chapter 26: Mesial temporal lobe epilepsy in children</b>	
Pavel Kršek, Michael Duchowny, Arthur Cukiert	305
Pathological substrates of pediatric temporal lobe epilepsy	305
Risk factors for pediatric temporal lobe epilepsy	305
Role of dual pathology in pediatric temporal lobe epilepsy	307
Clinical manifestations of temporal lobe epilepsy in children	309
EEG features of pediatric temporal lobe epilepsy	310
Surgical outcome in children with hippocampal sclerosis and dual pathology	310
<i>Timing of surgeries in relation to neuropsychological outcome</i>	310
<i>Surgical technique</i>	311
<i>Outcome regarding seizures</i>	311
<b>Chapter 27: Epilepsy surgery in MRI-negative patients</b>	
Thomas Bast, Philippe Kahane, Prasanna Jayakar	315
MRI-negative focal epilepsy	316
<i>Difference between MRI-negative and non-lesional epilepsy</i>	316
Selection of surgical candidates	318
Presurgical diagnostic work-up	319
Outcome after MRI-negative epilepsy surgery	324
<b>Chapter 28: Landau-Kleffner syndrome</b>	
Maria Clark, Rebecca Greenaway, Brian Neville	329
Background, clinical presentation and differential diagnosis	329
Multiple subpial transections of the dorsal surface of the temporal gyrus into the sylvian fissure	330
Mesial Subpial Transection indications	330
Recommended presurgical evaluation	331
Available results	331
Specific follow-up	332
New ideas about aetiology	333

## Section IV: SURGICAL TECHNIQUES FOR THE NEUROLOGIST

Editors: Gary Mathern, William D. Gaillard, Alexis Arzimanoglou

---

<b>Chapter 29: Lesionectomy</b>	
Thomas Blauwblomme, William Harkness, Christian Sainte-Rose	337
Patient selection	338
General principles	338
<i>Anesthesia protocol</i>	338
<i>Image guidance</i>	339
<i>Skin/bone/dura</i>	339
<i>Delineation of the lesion</i>	340
<i>Hemostasis/closure</i>	340
Safety	340
<i>Deep seated lesion</i>	340
<i>Eloquent areas</i>	340
Complete resection boundaries	342
<i>Lesion-specific considerations</i>	342
<i>Anatomic or electrophysiological limits?</i>	343

<i>Mesial temporal lobe lesions</i>	343
Respect of the non-epileptic brain	344
Future of lesionectomy?	344
<b>Chapter 30: Surgery for temporal lobe epilepsy</b>	
Johannes Schramm, Josef Zentner	347
Historical background of surgical treatment	347
Planning resection	348
General surgical remarks	348
Considerations in tumors associated with drug-resistant epilepsy	349
Resection strategies	349
<i>Surgical steps for standard anterior temporal lobectomy</i>	352
<i>Surgical steps for combined temporal pole and mesial resection</i>	353
<i>Surgical steps for transylvian selective amygdalo-hippocampectomy</i>	353
<i>Surgical steps for transcortical selective amygdalo-hippocampectomy</i>	354
<i>Surgical steps for subtemporal selective amygdalo-hippocampectomy</i>	354
<i>Surgical steps for extended lesionectomy</i>	355
Results	355
<i>Histopathological findings</i>	355
<i>Seizure outcome</i>	355
<i>Developmental and cognitive outcome</i>	356
<i>Operative complications</i>	357
<b>Chapter 31: Extratemporal localization and eloquent areas</b>	
Giorgio Lo Russo, Sanjiv Bhatia, Jeffrey G. Ojemann	361
Presurgical evaluation	361
Surgery	367
<i>Surgical decision-making</i>	367
<i>Anesthetic considerations</i>	367
<i>Neuronavigation and intra-operative MRI</i>	368
<i>Surgical technique: resection/disconnection</i>	368
<i>Multiple subpial transections</i>	372
<i>Intra-operative monitoring</i>	372
<i>Thermal ablation</i>	373
Outcomes	373
<i>Functional and developmental outcome</i>	374
<i>Complications</i>	375
<i>Pathology</i>	375
<b>Chapter 32: Corpus callosotomy: Surgical techniques</b>	
Bertil Rydenhag, Frank Ritter	381
Surgical history	382
Specific technical aspects, open surgery	382
Pitfalls	383
Results and outcome	384
Complication considerations	385
Considerations and caveats before performing the callosotomy	385
<b>Chapter 33: Hemispherotomy and multilobar surgery</b>	
Taisuke Otsuki	387
Historical background: from hemispherectomy to hemispherotomy	388
Description of the surgical procedures of hemispherotomy	389
Advantages and disadvantages of horizontal and vertical hemispherotomies	390
Multilobar disconnection and resection	391
Surgical planning	393
Perioperative management	393
Outcome	394
<b>Chapter 34: Surgical procedures for hypothalamic hamartomas</b>	
Georg Dorfmueller, Sarah Ferrand-Sorbets, Martine Fohlen, Olivier Delalande, John F. Kerrigan	399
The "classical" neurosurgical approaches	399
Recent alternative surgical techniques	400
<i>The transcallosal anterior interformiceal approach</i>	400
<i>The endoscopic approach</i>	400
<i>Stereotactic thermoablation</i>	405
<i>Gamma Knife Radiosurgery</i>	405
<i>Interstitial radiosurgery</i>	405
<i>Other surgical approaches</i>	406

<b>Chapter 35: Minimally invasive epilepsy surgery</b>	
Chima Oluigbo, Angus A. Wilfong	409
Principles and technological foundations of minimally invasive epilepsy surgery	410
Modifications of standard epilepsy procedures using smaller and “keyhole” craniotomies	411
<i>Minimally invasive approaches to temporal lobe epilepsy surgery</i>	411
<i>Minimally invasive approaches to hemispherectomy and other functional hemispherectomy variants of anatomical hemispherectomy</i>	412
Endoscopic epilepsy surgery	414
Endovascular epilepsy surgery	415
The future of minimally invasive epilepsy surgery	415
<b>Chapter 36: Pediatric epilepsy surgery techniques: a worldwide survey</b>	
Arthur Cukiert, Bertil Rydenhag, William Harkness	417
General issues	418
Medical intraoperative management	418
Skin preparation, opening and hemostasis	418
Closure	420
Postoperative imaging	420
Anesthetic agents	420
Temporal lobe resections	420
Callosal section	420
Hemispheric surgery	420
Temporal lobe tumors	422
Cavernoma	422
Complications	423
Neurosurgical pediatric epilepsy surgery training	423
<b>Chapter 37: Technical aspects of invasive monitoring</b>	
Jorge A. González-Martínez, Manfred Kudematsch	425
Localizing the epileptogenic zone	425
Localization of the functional/eloquent zone	426
The subdural method	426
The stereo-electroencephalography (SEEG) method	430
Invasive monitoring morbidity	432

## Section V. PALLIATIVE EPILEPSY SURGERY

Editors: Prasanna Jayakar, Gary Mathern, Alexis Arzimanoglou

---

<b>Chapter 38: Corpus callosotomy: Indications and results</b>	
Frank Ritter, Bertil Rydenhag	437
Corpus callosotomy and seizure generalization	437
Pre-operative evaluation and indications	438
Lennox-Gastaut syndrome and callosotomy	442
Other scenarios for callosotomy	443
Cognitive, behavioral, psychosocial, and quality of life outcomes	443
Consequences of callosotomy	444
<b>Chapter 39: Vagus nerve stimulation</b>	
Jo Sourbron, Lieven Lagae	447
VNS efficacy	448
Quality of life	451
Safety	452
<b>Chapter 40: Deep brain stimulation</b>	
Kristl Vonck, Sofie Carrette, Eric Kossoff, Paul Boon	455
History of deep brain stimulation in epilepsy	455
Overview of the different stimulation targets	456
<i>Epileptogenic network targets</i>	456
<i>Ictal onset zone DBS</i>	459
Closed-loop DBS	460
Deep brain stimulation in children	462

## Section VI. FOLLOWING SURGERY

### Editors: Alexis Arzimanoglou, J. Helen Cross

<b>Chapter 41: Long-term follow-up and transition to adult life</b>	
Douglas Nordli, Michael Duchowny, J. Helen Cross	471
Acute post-surgical care	471
Intermediate care	472
Long term follow-up and transition to adult life	473
<b>Chapter 42: Surgical failure, early recurrence and re-operation</b>	
Michael Duchowny, Douglas Nordli	475
Surgical failure	475
Reasons for failure: incomplete resection of the epileptogenic zone	475
<i>Incomplete pre-operative data</i>	476
<i>Co-localization of eloquent and epileptogenic cortex</i>	476
<i>Non-contiguous epileptogenic zones</i>	476
<i>Insufficient intracranial electrode coverage</i>	477
<i>Surgical inaccessibility</i>	477
Emergence of a new epileptogenic focus	477
Re-operation	478
<b>Chapter 43: Postoperative imaging</b>	
Chima Oluigbo, William D. Gaillard	485
Structural imaging modalities in the immediate and late postoperative period	486
Expected and unexpected brain imaging findings in the immediate postoperative period	487
Imaging and localization of implanted depth and subdural electrodes for invasive monitoring	489
Brain imaging as a research tool in the postoperative epilepsy patient	489
MRI safety of implanted neuromodulatory devices for epilepsy	490
<b>Chapter 44: Time to stop antiepileptic drugs</b>	
Kees Braun, Kim Boshuisen, Shlomo Shinnar	493
Why should we consider reducing AEDs?	493
What are the risks of reducing AEDs?	494
Do we know if postoperative AED reduction increases relapse risk?	495
What are the consequences of a seizure relapse following AED reduction?	496
In whom should we consider AED withdrawal?	496
When can we start to reduce AEDs?	497
<b>Chapter 45: Cognition, language, and memory outcomes</b>	
Mary Lou Smith, Torsten Baldeweg	503
Temporal lobe resection	503
Frontal lobe resection	506
Parietal or occipital lobe resection	507
Multilobar resection (excluding hemidisconnections)	508
Hemispheric resection	508
Age at surgery	510
Long-term follow-up	511
<b>Chapter 46: Psychosocial outcomes following seizure surgery</b>	
David Dunn, Jay Salpekar	515
Adult reports of psychosocial outcome	515
Social competence	518
Quality of life, family, and satisfaction with outcome	519
<b>Chapter 47: Comprehensive evaluation of outcomes</b>	
Brian Neville, J. Helen Cross	523
What is needed to report outcome?	523
Seizure outcome	523
Complications/unexpected events	525
Neurocognitive and psychiatric outcome	526
Psychosocial/quality of life	529

## Section VII. FUTURE PERSPECTIVES

---

### Chapter 48: A comprehensive epilepsy surgery program for children: State-of-the-art and future perspectives

Alexis Arzimanoglou, Philippe Kahane, J. Helen Cross, William D. Gaillard,  
Hans Holthausen, Prasanna Jayakar, Gary Mathern

	533
Specificities of childhood epilepsies	533
The surgical treatment gap	534
Candidates for a presurgical evaluation or a patient eligible for surgery: On what do they differ	535
Professional expertise	537
Which presurgical evaluation strategy and for whom	539
<i>Minimal requirements</i>	539
<i>Video-EEG recording of seizures</i>	541
<i>Other investigation tools</i>	541
<i>Invasive VEEG monitoring</i>	542
Postoperative long-term follow-up facilities	542
Future challenges and perspectives	542