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### Summary

Decision makers involved in the field of epilepsy surgery face a large event of ethical challenges. In particular, the establishment of outcome measures significant to the patient are crucial. Despite important advances in pre-surgical evaluations, these risks and benefits remain difficult to precisely stratify in individual cases. Truthful communication and an ethical attitude with the patient regarding the objective risk-benefit balance and the amount of medical uncertainty is warranted. Three illustrative situations are described and commented from an ethical perspective.

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**Keywords:** Epilepsy, surgery, ethics

### Aspects éthiques en chirurgie de l'épilepsie

Les personnes en charge du domaine de la chirurgie de l'épilepsie sont confrontées à un large éventail de défis éthiques. L'identification d'éléments pronostiques propres à chaque patient est en effet cruciale. Malgré l'importance des avancées dans les évaluations pré-chirurgicales, ces risques et bénéfices restent difficiles à stratifier de façon précise sur une base individuelle. Une communication basée sur la confiance, une attitude éthique face au patient avec mesure objective du rapport coûts-bénéfices et du degré d'incertitude médicale sont nécessaires.

Trois situations qui illustrent ces aspects sont présentées et commentées sous l'angle des considérations éthiques qu'elles soulèvent.

**Mots clés :** Epilepsie, chirurgie, éthique

### Ethische Fragen bei Epilepsiechirurgie

Entscheidungsträger im Bereich der Epilepsiechirurgie begegnen einer Reihe von ethischen Herausforderungen. Insbesondere ist es unabdingbar, bei jedem Patienten die prognostischen Faktoren zu identifizieren. Trotz grosser Fortschritte in der Epilepsiechirurgie bleibt es schwierig, die individuellen Risiken und Chancen eines Eingriffs bei einem bestimmten Fall genau zu bestimmen. Ehrliche Kommunikation und eine ethische Grundhaltung dem Patienten gegenüber sind bezüglich Risiko – Chancen-Abwägung und der verbleibenden medizinischen Unwägbarkeiten Bedingung. In diesem Artikel werden drei Situationen geschildert und aus ethischer Sicht kommentiert.

**Schlüsselwörter:** Epilepsie, Chirurgie, Ethik

### Introduction

Clinicians and decision makers involved in the field of epilepsy surgery face a large event of ethical challenges. Public health collective issues are naturally at play, and include the availability of resources, the identification and selection of potential candidates, waiting time to evaluation or surgery in a potentially deadly condition. On an individual patient level, medical decisions are taken in the context of an elective procedure that might lead to an immense improvement of quality of life but with unavoidable risks of major complications and often some long lasting deficits. The establishment of outcome measures significant to the patient (e.g. seizure freedom vs global quality of life) are crucial, both at individual level and at population level to assess the efficiency of the therapy. Despite important advances in non-invasive and invasive evaluations, these risks remain difficult to precisely stratify in individual cases. Truthful communication and an ethical attitude with the patient regarding the objective risk-

benefit balance and the amount of medical uncertainty is warranted, particularly with persons who often show cognitive, psychiatric and social comorbidities. In this article, we focus on individual aspects of ethical issues surrounding medical counselling before surgical intervention. Three illustrative situations are described and commented from an ethical perspective.

## Case 1

Mr X is a 28 year-old patient referred for presurgical evaluation. Since the age of 18, he suffers from sensory-motor seizures with occasional secondary tonic-clonic generalization. Seizures start with a stereotyped painful feeling in the left leg followed by tonic contraction of the trunk and four limbs. Occasionally, the seizure evolves towards version of the eyes and head to the left followed by tonic-clonic generalization. For the past 4 years, seizures have occurred exclusively during sleep, but have also occurred during wakefulness in the beginning of the disease, causing falls on each occasion. Several interictal EEG recordings have been unremarkable and MRI (3T with epilepsy-specific protocol) revealed no abnormality. Several drug treatments in monotherapy or polytherapy (carbamazepine, levetiracetam, lacosamide, lamotrigine) have failed to control the seizures or have led to side effects.

During the presurgical evaluation, typical seizures were recorded, whereas the ictal EEG only showed late midline rhythmic activity without interictal abnormalities. Interictal FDG-PET and ictal-interictal PET were non localising and imaging techniques based on electrophysiology (electric source imaging and simultaneous EEG-fMRI) were not possible given the absence of interictal spikes.

The conclusion was that the painful seizure onset in the left leg could be a manifestation from the primary or secondary somatosensory cortex or reflect a dystonic contraction related to premotor/supplementary motor involvement. In this context, invasive EEG was proposed to identify the epileptogenic zone.

As a clinician, you are convinced that the uncontrolled seizures with occasional generalizations are associated with increased morbidity and mortality (SUDEP). You think that intracranial EEG focusing on sensory and motor regions has a good chance of identifying the seizure onset but that close proximity or overlap with eloquent sensory or motor areas of the leg are likely, that could either prevent surgical resection after invasive recording or lead to a sensory or motor deficit. You anticipate a 2/3 chance of proceeding to surgery and a 50% chance of seizure freedom if surgical resection is performed. The risk for permanent deficit after electrode implantation is evaluated at <5%. Not knowing the seizure onset zone, prevents you from counselling about the risk of post-operative deficit. You want to balance these numbers with the <5% chance of being

seizure free with continuing best medical therapy and a mortality risk of 0.5-1% per year.

The patient is married and father of 3 young children, drives a car (he has only had nocturnal seizures for >3 years) and works as a skilled watchmaker. He is well aware of the risk of ongoing seizures but anxious about the risks of neurological deficit in case of an invasive procedure (implantation of electrodes and subsequent surgery). In your opinion, he exaggerates the risks and minimizes the chances of a good outcome.

## Commentary

How can you best advise this patient regarding surgery, while acknowledging your inability to give him precise predictions of his status after an intervention?

If a patient is capable of decision-making, as we shall assume is the case here, then the question is how to balance two different, and potentially conflicting, concerns. The first is a concern that the patient should make a free and informed decision based on a truthful understanding of the situation. The second is a concern that the patient should accept the intervention: the clinician here is convinced that the intervention is a good idea and may feel that if the patient refuses this reflects an inaccurate understanding of the situation.

The first concern, that the patient should gain a truthful understanding of the situation, reflects the difficulty of telling the truth in a manner that actually helps patients to understand their situation accurately. Respect for autonomy and self-determination require assisting the patient in his understanding of the available options and their consequences [1]. Yet patients regularly do not obtain or understand all the information relevant to their decision [2, 3]. Situations of clinical uncertainty, moreover, present clinicians with added difficulty in disclosing information truthfully and in an understandable manner [4]. Rather than simple disclosure of information, in situations such as this one, truth-telling requires assisting the patient in reaching a truthful understanding of his situation [5]. This requires tailoring the message to the patient's own representations in a manner that is adapted to the physician's relationship to the patient [6] as well as to the cultural and institutional context [7].

In proceeding with this explanation, it can be tempting for the physician to steer the patient towards acceptance of the intervention, rather than towards a decision that is more truly the patient's own. To a degree, it has been argued that this is acceptable. Removing biases that could affect the decision, such a general preference for inaction or fear of hypothetical costs, can assist the patient in reaching his own decision rather than manipulate him towards the one the physician wants [8]. Manipulating the patient, for example by presenting data in a manner that exaggerates the benefits or hides risks, is on the other hand clearly unac-

ceptable.

The main issue here, then, is one of truthfulness. The goal should be to help the patient to attain it, and thus to reach a truly informed decision.

## Case 2

A 35 year-old lady is referred for pharmaco-resistant temporal lobe epilepsy. She suffers since the age of 12 from seizures starting with a rising epigastric sensation followed by altered consciousness for about one minute with semi-purposeful manual gestures and deambulation. She is fully amnesic of seizures and her family reports many more than she feels she had. In parallel she complains of worsening memory and executive impairment. She lives alone, on welfare. MRI shows right-sided hippocampal sclerosis and a recent long-term EEG shows multiple daily seizures. Three episodes of poor medical compliance and drug withdrawal due to her cognitive deficits have led to repeated occurrences of secondary convulsive status epilepticus with admission to emergency department, intubation and drug-induced coma in intensive care unit, with durable worsening of cognitive impairment. Moreover, she suffers from depression but with poor adherence to psychiatric follow-up treatment. The patient is partly anosognosic of her condition given the amnesia of the seizures and her mood disorder causes negative anticipation of surgical complications.

You are convinced that the epilepsy is pharmaco-resistant and that the situation is repetitively worsened by compliance issues, despite a maximised support from community nurses at home. For you, surgery is the only option in this chronic unsatisfactory management. It could offer a great improvement for the patient and for the use of resources. You estimate that the risks of temporal lobe resection (memory decline, visual field deficit) are largely outbalanced by the potential benefits (risk of seizures/status epilepticus, iatrogenic complications of repetitive emergency admissions).

## Commentary

How can you best advise this patient, and how should a decision be reached in her case?

Here too, it can be tempting to attempt to convince the patient to accept the intervention, perhaps even soliciting her family's help in doing so. In a situation such as this one, however, where the patient seems anosognosic of her condition, the first question to address is that of decision-making capacity.

Adult patients are presumed to be competent and able to make their own decisions. Decision-making capacity, however, implies a number of abilities that are not present in all cases. When patients do not sufficiently possess these abilities, then we recognize that

they should sometimes be protected from their own troubled judgment. Decisions will then be made on other grounds than on their currently stated choices [9].

Formally assessing a person's decision-making capacity involves evaluating [10]:

- her understanding of the relevant information,
- her appreciation of the significance of this information for her circumstances,
- her ability to reason with the relevant information and weigh options logically, and
- her ability to express a choice.

Because each of these abilities will exist – or not – as regards choice-specific information, decision-making capacity must be assessed as regards a specific choice at a specific time. These abilities are not required to be perfect. Each should be present to a sufficient degree to consider that a person is capable of making a choice that is truly hers. In determining the necessary threshold of understanding and reasoning ability, clinicians should consider the degree of risk to the patient if her wishes are followed [9]. The higher the risk, the more demanding this threshold can be, but nevertheless perfection will still not be required.

When a patient's assessment of risks and benefits differs from that of the clinical team, as in the previous case, it is correct to ascertain whether or not he has understood the facts. Neither lack of agreement with the team nor even lack of understanding is sufficient to declare a patient incompetent. A patient must show not only lack of understanding but also the inability to understand in order to be considered incapable of decision-making.

In this case, then, the goal cannot be to convince the patient at all costs. Instead, the goal should be to ascertain whether or not she is capable of decision-making, in order to make the final decision appropriately. If she is capable of decision-making, then the purpose will be to help her to come to an informed decision even though she has no subjective experience of her seizures. If she is not capable of decision-making, then a decision must be made based on her previously stated wishes, if they are known, or on her best interests. The purpose would then be to keep the decision centred on the patient's own interests, despite the presence of third party interests (such as her family's, nurses', or social services') that may potentially come into conflict with her own.

### Case 3

This previously healthy boy presented with focal seizures at the age of 4 years. These initial events were reported as paroxysmal painful sensations in the eyes. Numerous clinical and subclinical seizures were recorded on EEG and were demonstrated as originating from the right parieto-occipital region. The initial MRI was normal. The frequency of seizures, initially rare and irregular, progressively increased to become refractory to various medical treatments, including valproic acid, vigabatrin, carbamazepine, oxcarbazepine, lamotrigine, clobazam and topiramate. At 6 years, motor seizures involving the left body side were observed, which were still noted on a daily basis one year later. The EEG showed an extended involvement of the right hemisphere, with the presence of numerous focal seizures still starting from the posterior regions, but followed by rapid generalization, and post-ictal slowing and attenuation in the ipsilateral frontal lobe. The boy also exhibited learning difficulties at school, but his neurological examination and repeated cerebral MRI remained normal at that time. A complete pre-surgical evaluation was proposed to the parents, who privileged the option of trying further medications before taking any risks linked to a potential surgical procedure. At 8 years, a third MRI showed a mild cortico-subcortical atrophy and signal abnormalities on FLAIR sequences involving the entire right hemisphere, consistent with the diagnosis of Rasmussen encephalitis. It is only one year later, at 9 years, that for the first time a discrete weakness in the left leg was noted on examination. The boy also had developed *epilepsia partialis continua*, with continuous seizures involving the entire left side of his body. His school difficulties had markedly increased, and the boy had been transferred to a specialized school for children with cognitive and motor problems. At that time, given that unfavorable evolution, the parents accepted a presurgical evaluation, which confirmed the diagnosis of Rasmussen encephalitis, all of the exams included in the work-up being concordant for a diffuse involvement of the right hemisphere and pointing to an unavoidable functional hemispherotomy. The boy was finally operated a few months later, at 11 years (i.e. 7 years after seizure onset), after two status epilepticus episodes that needed acute management in the intensive care unit. The intervention went without complications, but was followed by a worsening of the boy's hemiplegia and hemianopsia that had been diagnosed previously. The patient has remained seizure-free since, but he is unfortunately entirely dependent from caregivers in his daily activities. He is currently aged 22 years and lives with his mother.

A cognitive deterioration directly caused by refractory seizures, and in particular by a contralateral effect on the "healthy" hemisphere is strongly suspected in that disease. Likewise, a potential cognitive improvement can be expected if seizures are controlled.

### Commentary

Considering the fact that the surgical approach adopted in this case is the only curative option in Rasmussen encephalitis, but inevitably leads to motor and visual sequelae, how would you approach the ethical aspects that this patient's history illustrates? In particular, the question is whether such children should be considered for surgical intervention before they develop cognitive and motor difficulties that are very likely to appear at some point during the course of the disease.

One clear difficulty in this case is that a decision will have to be made one way or the other for a child who is not yet able to form his own decision. In other circumstances where life-altering decisions can be made during childhood, or deferred until the child is old enough to reach her own decision, the recommendation is usually to wait for this to happen while remaining aware that decision-making capacity can be attained before adulthood. Examples include sex-assignment surgery in children born with ambiguous genitalia [11], or genetic testing in the case of at-risk families [12, 13]. In the case of Rasmussen encephalitis, however, it is highly unlikely that this will be an option as cognitive decline usually sets in before a child is old enough to make his own decision.

Others, then, will need to step in and make this decision in the best interest of the child. In such situations, parents are usually the primary proxy for young children. They are expected to protect their child's best interests and make decisions based on this concern. Difficulties arise when parents are unable to provide such protection [14], or when parents' assessment of their child's best interest differs from that of health care providers. In such cases, centering the decision-making process on what constitutes a benefit for the child, what constitutes harm, and what actions are likely to bring about benefits and avoid harms, will often result in consensus. More difficult cases are sometimes arbitrated in court, as only judges can authorize clinicians to proceed despite lack of parental consent for a procedure. Even in such cases, however, the same criteria are applied and they raise similar difficulties [15].

In the present case, what constitutes the best interest of the child may be one of the central difficulties in the case. It is plausible to consider that avoiding cognitive decline is sufficiently important to warrant trading off on the risk of other forms of neurological deficits. Cognitive ability, after all, is valued as one of the central components of the human experience, and it also constitutes one of the means through which persons cope with the vagaries of life, including disabilities. The central issue here, then, will be to determine how this trade-off should be handled to respect the best interests of the child.

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