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REVIEW

The uncommon causes of status epilepticus: A Systematic Review

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Summary This paper reports the first systematic review of uncommon causes of status epilepticus reported in the literature between 1990 and 2008. Uncommon causes are defined as those not listed in the main epidemiological studies of status epilepticus. 181 causes were identified. These were easily categorised into 5 specific aetiological categories: immunological disorders, mitochondrial disorders, infectious diseases, genetic disorders and drugs/toxins. A sixth category of 'other causes' has also been included. Knowledge of these causes is important for clinical management and treatment, and also for a better understanding of the pathophysiology of status epilepticus.

Contents

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Introduction

Status epilepticus (SE) is a devastating neurological condition with a high morbidity and mortality if not treated immediately, with the mortality ranging from 7.6% to 43% (Chin et al., 2004).

The aetiology of SE plays an important role in determining its prognosis (Neligan and Shorvon, 2010; Rossetti et al., 2006) While the common causes of SE have been extensively studied (DeLorenzo et al., 1996; Wu et al., 2002)., the same cannot be said for uncommon causes. In large hospital or population based studies, the lists of uncommon causes (typically causes occurring at a frequency of <1%) are not presented, and the literature typically includes only case reports or small case series.

In this study, we aimed: (i) to identify all uncommon causes or uncommon conditions in which status epilepticus has been reported in the literature over a 20 year period; (ii) to categorise these; (iii) to assess where possible the extent to which treatment aimed at the underlying cause will alleviate the SE; (iv) to assess to what extent a consideration of these causes throws light on the mechanisms underlying the precipitation of status epilepticus. A brief summary of the some of the conditions in which SE is a particularly prominent or characteristic feature is also given.

Search strategy and selection criteria

We defined SE as ''an acute epileptic condition characterized by continuous seizures (partial or generalized, convulsive or nonconvulsive) for at least 30 min, or by 30 min of intermittent seizures without full recovery of consciousness between seizures'' (Wasterlain & Chen, 2006). In this study, we have defined an 'uncommon cause' as 'a cause of SE not reported (or not included in a separate category because they were so rare, typically <1% of causes) in the major epidemiological studies of SE (Chin et al., 2006; Coeytaux et al., 2000; DeLorenzo et al., 1996; Hesdorffer et al., 1998; Knake et al., 2001; Vignatelli et al., 2003; Wu et al., 2002). We chose this essentially operational definition in view of the absence of any incidence or prevalence figures on most of the conditions identified. Thus, 'uncommon causes' are defined as those other than:

1. Cerebrovascular diseases.
2. CNS infections.
   a. typical bacterial meningitis;
   b. viral encephalitis including JE encephalitis, herpes simplex encephalitis, Human Herpes virus 6;
   c. cerebral toxoplasmosis;
   d. tuberculosis and
   e. neurocysticercosis.
3. Intracranial tumours (both primary and secondary, benign or malignant).
4. Head trauma.
5. Alcohol related.
6. Withdrawal of or low levels of antiepileptic drugs.
7. Hypoxia/anoxia related.
8. Metabolic disturbances (electrolyte imbalances, glucose imbalance, organ failures, acidosis).

We identified relevant papers for this review by searches on PubMed, MEDLINE and Web of Science using the search terms ''status epilepticus'', ''epilepsia partialis continua'', and ''causes'', ''aetiology'', ''uncommon'', ''rare'' between 1990 and 2008. Articles were also identified through searches of the authors' own files. Only papers published in English were reviewed. Papers accepted were confined to those fulfilling the definition of 'uncommon causes'.

A total of 1931 articles were identified from PubMed. Of these, 56 articles were excluded as they were animal studies; 332 review articles were excluded; 986 articles were not relevant to the subject; 105 articles described cases with common causes. Thus a total of 450 articles were accepted, with a further 61 accepted articles identified through cross-referencing. A total of 300 articles were identified from Web of Science database. However, only 2 additional articles were accepted. Thus in total, 513 articles were accepted for this study. In some of these papers, 'status epileptics' was not defined (for instance in terms of duration), and in such papers we accepted the diagnosis met our criteria.

Results

181 uncommon causes of SE (defined above) were identified from the search in 588 articles reviewed. It proved

Table 1 Autoimmune disorders causing SE.

1. Paraneoplastic encephalitis
2. Hashimoto's encephalopathy
3. Anti-NMDA receptor encephalitis
4. Anti-VGKC receptor encephalitis
5. Rasmussen encephalitis
6. Cerebral lupus
7. Adult onset Still's disease
8. Anti GAD antibody associated encephalitis
9. Goodpasture syndrome
10. Thrombotic thrombocytopenic purpura
11. Antibody negative limbic encephalitis

References (principal references in bold): 1. (**Shavit et al., 1999**; Kinirons et al., 2006; Weimer et al., 2008; Koide et al., 2007), 2. (**Ferlazzo et al., 2006**; Janes et al., 2004; Striano et al., 2006; Tsai et al., 2007), 3. (**Dalmau et al., 2008**), 4. (Vincent et al., 2004), 5. (Capovilla et al., 1997), 6. (Tsuji et al., 2005), 7. (Hong and Lee, 2008), 8. (Kanter et al., 2008), 9. (Rydel and Rodby, 1998), 10. (Blum and Drislane, 1996), 11. (Samarasekera et al., 2007).

Table 2 Mitochondrial diseases causing SE.

1. Alpers disease
2. Occipital lobe epilepsy/MSCAE
3. Mitochondrial encephalopathy, lactic acidosis and stroke-like episodes (MELAS)
4. Leigh syndrome
5. Myoclonic encephalopathy with ragged red fibres (MERRF)
6. Neuropathy, ataxia and retinitis pigmentosa (NARP)

References (principal references in bold): 1. (**Naviaux and Nguyen, 2005**), 2. (**Tzoulis et al., 2006**), 3. (**Huang et al., 2002**; **Goto et al., 1990**; **Crimmins et al., 1993**; **Leff et al., 1998**; **Alemdar et al., 2007**) 4. (**Elia et al., 1996**), 5. (Sitburana et al., 2001), 6. (Keranen and Kuusisto, 2006).

possible to place these into 5 categorical groups — immunological disorders (Table 1), mitochondrial disorders (Table 2), infectious diseases (Tables 3 and 4), genetic disorders (Table 5), drugs and toxins (Table 6), and also a final group comprising causes not listed in the first 5 categories (Table 7).

Immunological disorders

SE was an important symptom in a variety of different immunological disorders (Table 1).

Paraneoplastic encephalitis

Paraneoplastic encephalitis is a rare cause of SE. It occurs predominantly in adult females with the typical semiology being epilepsia partialis continua (EPC) or nonconvulsive SE (NCSE) (Kinirons et al., 2006; Koide et al., 2007; Shavit et al., 1999; Weimer et al., 2008; Yang et al., 2006). Other common clinical features include dysautonomic features, palatal and tongue myoclonus, peripheral neuropathy and cerebellar signs. Seizures may precede the onset of SE (Weimer et al., 2008).

Control of SE relies on treatment of the underlying aetiology with chemotherapy (Shavit et al., 1999; Kinirons et al., 2006), tumour resection (Weimer et al., 2008; Yang et al., 2006) or a combination of both (Koide et al., 2007).

Hashimoto's encephalopathy

Reports of cases of SE in Hashimoto's encephalopathy were mainly in adult females (Ferlazzo et al., 2006; Janes et al., 2004; Striano et al., 2006; Tsai et al., 2007). A prior history of seizures was noted in 2 cases (Striano et al., 2006; Tsai et al., 2007). Generalized convulsive SE (GCSE) was the commonest form of SE, while recurrent SE was rarely reported (Ferlazzo et al., 2006). Cases were either euthyroid or hypothyroid.

Control of SE was poor with antiepileptic drugs, while aggressive steroid therapy using intravenous methylprednisolone for 3—5 days followed by oral steroids produced dramatic recovery. Death has been reported due to refractory status (Striano et al., 2006).

Anti-NMDA receptor encephalitis

Anti-NMDA-receptor encephalitis affects predominantly young females (91%; median age 23 years) and may be associated with an underlying ovarian tumour (53%) (Dalmau et al., 2008). Starting with a prodrome of headache, low-grade fever, or a non-specific viral-like illness, it progresses to prominent neuropsychiatric symptoms, seizures, dyskinesias and later central hypoventilation and autonomic instability. SE occurred in 8 of the 100 cases identified. The condition is treatment responsive — 75% of patients recovered with 47% achieving full recovery, although a characteristic feature during recovery is a persisting amnesia of the disease process.

Treatment included tumour resection or immunotherapy (steroids, immunoglobulins, plasma exchange or chemotherapy with cyclophosphamide or rituximab) and the response to treatment appeared to be associated with the timing of tumour diagnosis.

Mitochondrial disorders

SE is a prominent feature of a number of uncommon mitochondrial disorders (Table 2).

POLG1 mutations

POLG1 mutations have recently gained much attention and to date, 2 clinical syndromes have been associated with SE. Alpers disease, a disease of early childhood and only recently linked with POLG1 mutations (Naviaux and Nguyen, 2005), features focal SE (EPC or myoclonic) as its usual presenting symptom, preceded by an infectious illness and ending with refractory seizures. Other clinical features include episodic psychomotor regression and liver failure. The mean duration from time of presentation to death is 12 months, with liver failure as the fatal event (Narkewicz et al., 1991; Nguyen et al., 2005).

The second clinical syndrome is associated with the A467T and W748S POLG1 mutation (Tzoulis et al., 2006). Both epilepsy and SE are common features, whereby there was a predilection for occipital seizure phenomena includ-

Table 3 Infectious disease as uncommon causes of SE (see Supplementary Table 1 for additional references).

Atypical bacterial infections	*Viral infections*
1. Bartonella/Cat-scratch disease	1. HIV and HIV related infections
2. Neurosyphilis	2. Arboviruses
3. Coxiella Burnetti (Q fever)	3. Progressive multifocal leukoencephalopathy (JC virus)
4. Mycoplasma pneumonia	4. Parvovirus B19
5. Scrub typhus	5. Varicella encephalitis
6. Shigellosis	6. SSPE
7. Chlamydophila psittaci	7. Measles encephalitis
Prion disease	8. Rubella encephalitis
1. Creutzfeldt-Jakob disease	9. RSV associated SE
Other infections	10. Polioencephalomyelitis
1. Paracoccidioidomycosis	
2. Paragonimiasis	
3. Mucormycosis	

Selected references (principal references in bold): *Atypical bacterial infections*: 1. (**Armengol and Hendley, 1999**; Carithers and Margileth, 1991), 2. (**LoVecchio, 1995**; Gürses et al., 2007), 3. (Ravid et al., 2008), 4. (Jeffery et al., 1995); *Prion disease*: 1. (**Rees et al., 1999**; Neufeld et al., 2003); *Viral infections*: 1. (**Wong et al., 1990**; Van Paesschen et al., 1995; Lee et al., 2005), 2. (**Bosanko et al., 2003**), 3. (Berciano et al., 2003), 4. Erol et al., 2006; *Other infections*: 1. (Franca et al., 2005).

ing flickering lights, oculoclonus, dysmorphopsia, micro- or macropsia and pallinopsia (Engelsen et al., 2008). The semiology of SE includes NCSE, convulsive SE and simple partial SE. Other clinical features consisted of a combination of ataxia, headaches, peripheral neuropathy, nystagmus, myoclonus, opthalmoplegia and liver involvement.

There is no effective therapy and the severity of epilepsy appears to correlate closely with disease morbidity and mortality with deaths due to treatment resistant SE or hepatic failure. Valproate induced liver failure was noted in 5 cases (Tzoulis et al., 2006).

MELAS

MELAS (mitochondrial myopathy, encephalopathy, lactic acidosis, stroke-like episode) is a mitochondrial disorder frequently due to the A3243G point mutation (Goto et al., 1990; Huang et al., 2002). MELAS may present with any forms of SE, including NCSE (Crimmins et al., 1993; Feddersen et al., 2003; Leff et al., 1998), SPSE (Nakamura et al., 2000), or GCSE (Alemdar et al., 2007; Liou et al., 2000). Associated clinical features were due to the underlying disease — these include hemiparesis and hemianopia due to stroke-like episodes, sensorineural deafness, migraine, speech or visual disturbances (Alemdar et al., 2007; Crimmins et al., 1993; Feddersen et al., 2003; Leff et al., 1998).

With the exception of cases of NCSE which showed good response to standard AED treatment, prognosis is generally poor. Neurological sequelae noted include dysphasia, cortical blindness, hemiparesis, myoclonus, dementia, deafness and Anton-Babinski syndrome (Alemdar et al., 2007; Leff et al., 1998; Peterus et al., 1997). Mortality was reported in 7 cases — and in one series, all 4 cases died as a result of SE (Huang et al., 2002).

Infectious diseases

SE is a prominent feature of a number of less common infectious diseases (Table 3), in addition to those listed in the major epidemiological studies.

Cat-scratch disease encephalopathy

Cat-scratch disease (CSD) is a seasonal infection affecting children and caused by a pleomorphic gram-negative bacillus, *Bartonella henselae* (Goral et al., 1994). Between 0.17% and 2% of cases manifests as CSD encephalopathy (Carithers and Margileth, 1991). SE associated with CSD encephalopathy is predominantly GCSE. Common clinical features include fever, regional lymphadenopathy and skin excoriations. (Armengol and Hendley, 1999) while uncommon features include transient postictal combative behaviour, hemiparesis, tremor and chorea (Hadley et al., 1995; Yagupsky and Sofer, 1990).

Treatments given include antiepileptic medications, antibiotics (Ceftriaxone, Gentamicin or Amoxycillin) and acyclovir. 2 cases were refractory to antiepileptic drugs (Hahn et al., 1994; Yagupsky and Sofer, 1990), while 1 case was given steroids and mannitoll (Ashkenasi et al., 1993). All cases recovered from their illnesses; partial epilepsy was reported as a neurological sequelae (Hahn et al., 1994).

HIV and HIV related disorders

New onset seizures have been reported in 4—11% of HIV seropositive patients, and of these, 8.1—18% develop SE (Lee et al., 2005; Van Paesschen et al., 1995; Wong et al., 1990). Both seizures and SE may be due to a variety of causes (Table 4). The commonest SE semiology is GCSE, occurring in 62.5—100% of cases (Van Paesschen et al., 1995). NCSE has also been reported (Wong et al., 1992) while EPC usually occurs in association with concurrent progressive multifocal leukoencephalopathy (PML) (Bartolomei et al., 1999; Ferrari et al., 1998). Concurrent hypomagnesemia and renal failure appears to increase the risks of convulsive SE (Van Paesschen et al., 1995).

SE was responsive to benzodiazepine treatment with or without the addition of phenytoin (Lee et al., 2005). Low average CD4 counts and the duration of SE are poor prognostic factors associated with high mortality (Lee et al., 2005).

Table 4 Aetiology of SE in patients with HIV.

Opportunistic CNS infections
(Toxoplasma infections, cryptococcal meningitis, cytomegalovirus infections, HSV infections, other infections)
Associated structural lesions
CNS tumours (lymphoma), acute stroke, CNS trauma
Metabolic abnormality
(Hyponatremia, hypomagnesemia, hypocalcemia, renal impairment)
Alcohol withdrawal
Drug related
Antiretroviral drugs toxicity, withdrawal of AEDs
Non-specific HIV disease (primary HIV infection)

References: (Lee et al., 2005; Van Paesschen et al., 1995).

Creutzfeldt-Jakob disease

Creutzfeldt-Jakob disease is a disease of mutated prion proteins. A rapidly progressive dementia with behavioural abnormalities, cerebellar dysfunction and a typical myoclonus constitutes its common presentation. SE is an uncommon presentation, mostly associated with the sporadic form of CJD and tends to present late in the disease process (Neufeld et al., 2003; Rees et al., 1999). The majority of cases have been GCSE or NCSE (Cokgor et al., 1999; Neufeld et al., 2003; Rees et al., 1999) with EPC rarely reported (Parry et al., 2001).

Cortical or subcortical signal changes as well as cerebral atrophy have been noted on MRI studies. EEG findings include the characteristic periodic sharp waves (PSWs), pseudoPSWs, PLEDs, variable sharp waves and in 1 case, stimulus-induced rhythmic periodic or ictal discharges (SIRPIDS) (Rossetti and Dunand, 2007).

SE was universally refractory to AED therapy, and time from SE presentation to death ranged from 2 weeks to 3.5 months (Cokgor et al., 1999; Neufeld et al., 2003; Parry et al., 2001; Rees et al., 1999).

Genetic disorders

SE is a common feature of a number of rare genetic disorders (Table 5).

Ring chromosome 20

Ring chromosome 20 is a rare condition arising from deletion of its long arm, where 2 important epileptogenically associated genes are located, or from disordered equilibrium of the remaining genes (Biraben et al., 2004; Inoue et al., 1997). Typically affecting children between 2 and 14 years, it presents with NCSE, simple motor seizures, automatisms, low IQ with behavioural regression, and occasional dysmorphism.

Most cases are refractory to treatment with AEDs. Despite this, only 1 death has been reported due to prolonged seizures from this condition (Jacobs et al., 2008).

Angelman syndrome

Of the 5 known genetic defects associated with Angelman syndrome, only cases with *de novo* deletions have reported occurrences of SE (Uemura et al., 2005; Valente et al., 2006) It is a common phenomenon (33.3—90.1%) (Ohtsuka et al., 2005; Sugimoto et al., 1992), presenting in children between 13 and 24 months and (Valente et al., 2006; Laan et al., 1997) frequently manifests as an atypical absence SE.

The seizures tend to fizzle out with age and treatment, with seizure freedom achieved at 8 years in a Japanese series (Ohtsuka et al., 2005). VPA and clonazepam were reported to be the most effective AEDs (Galvan-Manso et al., 2005). Other clinical aspects of the disease, however, remain persistent and most cases eventually develop severe mental retardation, behavioural problems, and motor dysfunction.

Porphyria

The porphyrias are a group of metabolic disorders affecting the haeme biosynthesis pathway due to specific enzyme defects. 2 forms of porphyrias have been associated with SE — the acute intermittent porphyrias (AIP) and acute hepatic porphyrias (AHP) (Bhatia et al., 2008; Zaatreh, 2005). All reported cases were convulsive in semiology, 2 of which had an underlying history of epilepsy and recurrent SE. 1 patient developed refractory SE in pregnancy which ended following termination.

Treatment options are difficult as most first line antiepileptic drugs are proporphyrogenic (Bhatia et al., 2008). AEDs successfully used in treating SE are levetiracetam, gabapentin and propofol (Bhatia et al., 2008; Zaatreh, 2005).

Drugs/toxins

SE is also a prominent feature of a number of rare toxic exposures (Table 6).

Antiepileptic drugs

Tiagabine (TGB), a nipecotic acid analogue, has been implicated in the causation of status epilepticus (Kellinghaus et al., 2002) particularly nonconvulsive status epilepticus, even in patients without a prior diagnosis of epilepsy (Jette et al., 2006; Vollmar and Noachtar, 2007). It is thought to be pro-convulsant due to stimulation of presynaptic $GABA_B$ receptors which in turn inhibits the inhibitory postsynaptic potential mediated by $GABA_A$ receptors (Chan and Yung, 1999; Kellinghaus et al., 2002; Leikin et al., 2008). In a review of the safety data of TGB from clinical trials, the authors concluded that treatment with TGB at recommended doses did not increase the risk of status epilepticus in patients with partial seizures (Shinnar et al., 2001). However, Koepp et al. estimated the annual incidence of TGB induced NCSE at 6.7% per TGB treatment year and concluded that patients on TGB are at higher risks of developing NCSE compared to non-TGB users (Koepp et al., 2005). GCSE have also been reported and appears to be related to toxic ingestion (Leikin et al., 2008). The main EEG finding was generalized spike wave discharge (Koepp et al., 2005; Shinnar et al., 2001).

Clinical improvement was seen with a combination of either reduction or withdrawal or TGB with or without acute

Table 5 Status epilepticus due to genetic diseases (103) (see Supplementary Table 2 for additional references).

Chromosomal aberrations	*Malformations of cortical development*
1. Ring chromosome 20	1. Focal cortical dysplasias
2. Angelman syndrome	2. Hemimegalencephaly
3. Wolf-Hirshhorn syndrome	3. Polymicrogyria
4. Fragile X syndrome	4. Heterotopias
5. XLMR syndrome	5. Schizencephaly
6. Ring chromosome 17	*Neurocutaneous syndromes*
Inborn errors of metabolism	1. Sturge Weber syndrome
1. Porphyria	2. Tuberous sclerosis
2. Menkes disease	*Others*
3. Wilson's disease	1. Dravet syndrome
4. Adrenoleukodystrophy	2. Familial hemiplegic migraine
5. Alexnader disease	3. Progressive myoclonus epilepsies
6. Cobalamin C/D deficiency	4. Infantile onset SCA
7. OTC deficiency	5. Wrinkly skin syndrome
8. Hyperprolinaemia	6. Neurocutaneous melanomatosis
9. Maple syrup urine disease	7. Neuroserpin mutation
10. 3-Methylcrotonyl CoA carboxylase deficiency	8. Wolfram syndrome
11. Lysinuric protein intolerance	9. AR hyperekplexia
12. Hydroxyglutaric aciduria	10. Cockayne syndrome
13. Metachromatic leukodystrophy	11. CADASIL
14. Kuf's disease	12. Robinow syndrome
15. Beta ureidopropionase deficiency	13. LYK-5 mutation
16. 3 hydroxyaxyl CoA dehydrogenase deficiency	14. MECP2 mutation
17. Carnitine palmytoyltransferase deficiency	15. Malignant hyperpyrexia
18. Succinic semialdehyde dehydrogenase deficiency	

Selected references (principal references in bold): *Chromosomal aberrations*: 1. (**Biraben et al., 2004**; Inoue et al., 1997), 2. (Ohtsuka et al., 2005; Sugimoto et al., 1992), 3. (Battaglia et al., 1999); *Inborn errors of metabolism*: 1. (**Bhatia et al., 2008**; Zaatreh, 2005), 2. (**Bahi-Buisson et al., 2006**), 3. (**Turk-Boru et al., 2003**); *Malformations of cortical development*: 1. (**Fauser et al., 2006**); *Others*: 1. (Buoni et al., 2006), 2. (Beauvais et al., 2004), 3. (Kumada et al., 2006).

benzodiazepine therapy (Koepp et al., 2005; Shinnar et al., 2001; Vinton et al., 2005).

Carbamazepine may cause paradoxical NCSE (Marini et al., 2005) while in toxic doses, may result in convulsive SE associated with wide complex tachycardia, antiepileptic drug resistance and mortality (Sharma et al., 1992). Valproic acid (VPA) has been hypothesized to cause SE in rare instances via induced hepatotoxicity and hyperammonemia, hyperammonemia alone, or via a paradoxical effect involving GABA or excitatory amino acids (Capocchi et al., 1998). Most of the reported cases were NCSE, within 2 weeks of starting VPA or within a week of dose increase (Capocchi et al., 1998; Shahar et al., 2002).

Antimicrobials

Cephalosporins are epileptogenic due to its structural similarity with bicuculline, thus exerting an inhibitory effect on GABA receptors (De Sarro et al., 1995). Usage of Cefepime, Ceftazidime and Ceftriaxone have all been complicated by NCSE (Martinez-Rodriguez et al., 2001; Ozturk et al., 2009). In contrast, GCSE was seen with intrathecally administered Cefazoline together with iohexol (Choi et al., 2008). Renal impairment was a prominent feature, either as an end-stage event or as an acute renal failure (Martinez-Rodriguez et al., 2001; Ozturk et al., 2009). This was thought to be due to increased blood brain barrier penetration as a result of reduced plasma protein levels and albumin-antibiotics binding affinity in uraemia, as well as competitive inhibition by organic acids on antibiotic active transport (Chatellier et al., 2002; Wallace, 1997).

Recovery was reported in most cases occurring 1—7 days following cessation of cephalosporin and concurrent antiepileptic therapy.

Isoniazid toxicity is another frequently reported cause of SE (Caksen et al., 2003; Tajender and Saluja, 2006; Tibussek et al., 2006). It consists of a triad of coma, metabolic acidosis and refractory seizures preceded by nausea, vomiting, fever, rashes and ataxia. The mechanism of seizure production is due to isoniazid induced pyridoxine deficiency, and subsequent secondary reduction in GABA synthesis (Wood and Peesker, 1972).

Replacement of pyridoxine lead to termination of SE in all reported cases.

Chemotherapeutic drugs

Ifosfamide, a nitrogen mustard derivate, is a myelosuppressive agent from the group of alkylating compounds. Ifosfamide encephalopathy consists of ataxia, confusion, cerebellar signs, seizures, mutism, visual hallucinations and extrapyramidal features (Nicolao and Giometto, 2003). NCSE is a rare complication and presents with confusion, echolalia, mutism or myoclonus (Primavera et al., 2002; Wengs et al., 1993). The mechanism of action is thought to be derived from its active form, chloroacetaldehyde (CAA) being oxidised and conjugated to cysteine to form S-carboxymethylcysteine (SCMC), which has agonistic effects

Table 6 Drugs/toxins that cause or precipitate status epilepticus (see Supplementary Table 3 for additional references).

Antiepileptic drugs	*Antipsychotics*	*Chemotherapeutic drugs*
1. Tiagabine[a]	1. Olanzapine	1. Ifosfamide[a]
2. Carbamazepine	2. Sertindole	2. Cisplatin
3. Lamotrigine	*Contrast media*	3. Tacrolimus
4. Levetiracetam	1. Iohexol	4. Cyclosporin A
5. Valproic acid	2. Fluorescein	5. Methotrexate
6. Vigabatrin	3. Iopamidol	6. Etoposide
7. Gapapentin	4. Diatriazoate	*Others*
8. Pregabalin	*Toxins*	1. Theophylline[a]
9. Topiramate	1. Star fruit[a]	2. Lithium[a]
Antimicrobials and antiviral drugs	2. Endosulfan	3. Baclofen
1. Cephalosporins[a]	3. Domoic acid[a]	4. Morphine
2. Isoniazid[a]	4. Cocaine	5. Dramamine
3. Quinolones	5. Tetramine[a]	6. Flumazenil
4. Antiviral	6. Camphor	7. N-Acetylcysteine
5. Antihelmintic	7. Aluminium containing biomaterial	8. 4-Aminopyridine
6. Antimalarial	8. Carbon monoxide	9. Allopurinol
Antidepressants	9. Colloidal silver	10. Calcium carbonate
Bupropion[a]	10. Ecstacy	11. Clonidine
Amoxapine	11. Lead	12. Corticosteroids
Clomipramine	12. Lysergic acid amide	13. Gelatine
Fluoxetine	13. Maneb	14. Fligrastim/molgramostim
Amitriptyline	14. Neem oil	15. Interferon
Citalopram	15. Nitromethane	16. Mexiletine
Fluvoxamine	16. Petrol sniffing	17. Propofol
		18. Sulfasalazine
		19. Thyroxine

Selected references: *Antiepileptic drugs*: 1. (Koepp et al., 2005), 2. (Marini et al., 2005), *Antimicrobials*: 1. (Martinez-Rodriguez et al., 2001), 2. (Caksen et al., 2003); *Antidepressants*: 1. (Morazin et al., 2007); *Constrast media*: 1. (Tahta et al., 1993); *Toxins*: 1. (Neto et al., 2003), 2. (Grimmett et al., 1996), 3. (Teitelbaum et al., 1990); *Chemotherapeutic drugs*: 1. (Wengs et al., 1993); *Others*: 1. (Krieger and Takeyasu, 1999), 2. (Bellesi et al., 2006), 3. (Saltuari et al., 1992).

[a] Frequently reported causes of SE.

on the AMPA — kainite receptors (Nicolao and Giometto, 2003).

Response to therapy with standard AED therapy was favourable with full recovery.

Toxins

Domoic acid intoxication was first reported in 1987 in Canada following an outbreak of an encephalopathy consisting of headaches, hemiparesis, opthalmoplegia, seizures and altered consciousness, associated with ingestion of contaminated blue mussels (*Mytilus edulis*) (Perl et al., 1990; Teitelbaum et al., 1990). Domoic acid is a structural analogue of kainic acid, a powerful glutamate receptor agonist (Quilliam and Wright, 1989) and overactivation leads to seizure induction as well as neuronal damage (Debonnel et al., 1989). Seizures, myoclonus and GCSE have been reported following acute intoxication, and complicated by chronic memory deficits and rarely temporal lobe epilepsy (Cendes et al., 1995; Teitelbaum et al., 1990).

Star fruit (*Averrhoa carambola*) a tropical fruit originating from South East Asia, causes neurotoxicity among patients with chronic renal insufficiency (Chang et al., 2000; Neto et al., 2003). Neto reported three levels of intoxication with status epilepticus presenting as the most severe form. In his series of 32 patients with star fruit toxicity, 7 developed seizures and all but 1 died, mostly due to SE (Neto et al., 2003). No exact mechanism has been identified, although extracts from star fruit have been shown in animal studies to inhibit GABA binding on synaptic membranes (Carolino et al., 2005).

There is no specific treatment for toxin induced SE. Cases of SE due to camphor poisoning have been reported to respond to anticonvulsant therapy (Emery and Corban, 1999). However, success or failure of treatment appears to be related to the dose of toxins ingested. Apart from a few case reports showing success using multiple anticonvulsants to control SE secondary to endosulfan, domoic acid or star fruit poisoning (Grimmett et al., 1996; Chang et al., 2000), treatment is largely confined to supportive care with a high morbidity and mortality rate.

Other causes

The final table (Table 7) presents reported iatrogenic causes of SE such as electroconvulsive therapy (Povlsen et al., 2003), neurosurgical complications (Burneo et al., 2005), uncommon complications of common medical conditions like

Table 7 Other causes of SE (see Supplementary Table 4 for additional references).

Iatrogenic	*Other medical conditions (cont.)*
1. Electroconvulsive therapy	7. Ulcerative colitis
2. Temporal lobectomy	8. Behcet
3. Insertion of intracranial electrode	9. Coeliac disease
4. Ventriculoperitoneal shunt	10. Cobalamin deficiency
5. Blood transfusion	11. Folinic acid responsive seizures
6. Carotid angioplasty and stenting	12. Renal artery stenosis
Other medical conditions	13. Pituitary apoplexy
1. Multiple Sclerosis	14. Renal artery dissection
2. Hypertension induced PRES	15. Hypomelanosis if Ito
3. Neuroleptic malignant syndrome	16. Cerebral palsy
4. Panayiotopoulus syndrome	17. Hemophagocytic lymphohistiocytosis
5. Thyroid disease	18. Anhidrotic ectodermal dysplasia
6. Pyridoxine dependent seizure	19. Methaemoglobinaemia

Selected references: *Iatrogenic:* 1. (Povlsen et al., 2003), 2. (Burneo et al., 2005); *Other medical conditions*: 1. (Hess and Sethi, 1990), 2. (Al-Ansari and Todwal, 2007), 3. (Yoshino et al., 1998).

multiple sclerosis (Hess and Sethi, 1990) and hypertension (Al-Ansari and Todwal, 2007), in addition to a compilation of single case reports of rare causes of SE.

Conclusions

Knowledge of the range of conditions is obviously important to clinical practice and to diagnosis and investigation. Over 180 different aetiologies causing SE deemed uncommon were identified. Obviously, this sort of survey is open to reporting bias, and the list of causes cannot be claimed to be either complete or comprehensive. Nevertheless, it is difficult to see how else this topic could be approached, and interestingly most of the causes identified fell easily into 5 major aetiological groups (and a 6th miscellaneous category), suggesting that there may be common clinical attributes and pathophysiological mechanisms.

In treating status epilepticus it must be remembered that the associated mortality and morbidity is primarily due to the underlying disease or its complications rather than from a direct result of SE (Neligan and Shorvon, 2010) and aetiologies of SE associated with a particularly higher rate of mortality include Alpers disease, West Nile encephalitis, CJD and star fruit toxicity. An exception to this is the refractory nature of the SE in the POLG1 mutation associated syndromic epilepsy, where 9 of the 11 deaths were due to treatment resistant SE. Furthermore, SE often responds to therapy aimed at the underlying cause rather than symptomatic treatment of the seizures as is demonstrated by the success of immuno-suppressants in stopping SE in immunological disorders, the treatment of infections or the removal of toxic causes. Very often the success of a treatment strategy is dependent upon understanding the underlying aetiology.

The aetiologies identified in this review include conditions where SE is a frequent or predominant manifestation of the epileptic disorder (e.g. POLG1 mutations, ring chromosome 20, and domoic acid intoxication) and these conditions are of particular interest from the point of view of understanding the mechanisms of SE. Among the known or proposed pathophysiological mechanisms of SE due to the uncommon causes, 3 distinct groups stand out:

(1) Derangements of neurotransmitter function, in particular $GABA_A$ function — for instance, by agents such as tiagabine, valproic acid, vigabatrin, cephalosporins and endosulfan.
(2) An inflammatory process from autoimmune (e.g. paraneoplastic encephalitis, Hashimoto's encephalopathy, Rasmussen's encephalitis), infectious (e.g. CJD, CSD encephalopathy) and non-infectious entities (e.g. multiple sclerosis).
(3) Mitochondrial oxidative stress and mitochondrial DNA damage on epileptogenesis. Mitochondrial dysfunction may be the cause or consequence of epileptic seizures or SE (Cock, 2007; Patel, 2004).

It is our contention that by producing a preliminary and relevant list of the uncommon cause, it may encourage more awareness as well as reporting of similar conditions causing SE. An understanding of the cause is of obvious important in targeting investigation and improving prognosis. It may also lead to new insights into the mechanisms of the induction of status epilepticus and possibly also to better approaches to treatment.

Appendix A. Supplementary data

Supplementary data associated with this article can be found, in the online version, at doi:10.1016/j.eplepsyres.2010.07.015.

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