

Epidemiology of Idiopathic Generalized Epilepsies

Pierre Jallon and Patrick Latour

Epilepsy and EEG Unit, University Hospital, CH 1211, Geneva 14, Switzerland

Summary: Idiopathic generalized epilepsies (IGEs) are a relatively new category of disorders defined by strict clinical and electroencephalogram (EEG) features proposed by the International League Against Epilepsy (ILAE) classification of epileptic syndromes. IGEs are usually easy to diagnose when clinical and EEG data are collected, but epilepsy is not synonymous with epileptic syndrome. So far, IGEs are studied in the large group of epilepsies of undetermined or unknown etiology although the genetic origin is now largely accepted. ILAE-proposed criteria are helpful in the clinical and therapeutic management of IGEs, but many epidemiologic studies still confuse the cryptogenic and idiopathic groups. Some syndromes in childhood, which are completely described by strict electroclinical criteria such as the absence epilepsies, juvenile myoclonic epilepsies, are usually included and analyzed in epidemiologic studies; however, other epileptic syndromes observed in infancy, such as benign familial neonatal seizures and benign myoclonic epilepsy in infancy, are quite rare and are usually excluded from epidemiologic surveys because they are difficult to describe completely

in electro-clinical terms. Another strong limitation in the study of epidemiology of IGEs is the lack of EEG data, either because EEG is not available or the routine EEG is normal. This is particularly relevant in the inclusion of patients with only tonic-clonic seizures. IGEs encompass several different syndromes, and a few patients shift from one phenotype to another. The overlapping of some syndromes during infancy and adolescence increased the difficulty to individualize strictly the correct syndrome. Many discrepancies can be observed in the distribution of the different syndromes included in the group of IGEs, because the strict criteria for classifying these syndromes proposed by the ILAE are often not respected. With this understanding, the general frequency of IGEs can be assessed at 15–20% of all epilepsies. The frequency and the distribution of incidence and prevalence of the different syndromes are tentatively reported and discussed. When the term idiopathic is used following the restrictive ILAE criteria, the mortality data concerning patients with idiopathic epilepsies do not show an increased standardized mortality ratio.

Key Words: Epilepsy—Idiopathic epilepsies—Epidemiology.

Idiopathic generalized epilepsies (IGEs) are a relatively new category of disorders defined by strict clinical and electroencephalogram (EEG) features proposed by the International League Against Epilepsy (ILAE) classification of epileptic syndromes (1).

Epidemiologic data concerning IGEs are scarce or often unreliable because most descriptive studies did not specifically analyze these syndromes or because the investigations—mostly the EEG data—on which the diagnoses were based were not always available.

This review will be limited to epidemiologic data concerning the various well-defined IGE syndromes described with the restrictive ILAE criteria.

METHODOLOGY ISSUES

Definition of IGEs is a crucial point for the evaluation of its incidence and prevalence. The international classification of epileptic syndromes has proposed strict clinical

and EEG criteria to individualize IGEs. Clinical experience has shown that these criteria are not always fulfilled by every patient every time and that IGEs represent a heterogeneous condition in which many factors, such as age of onset, external factors, role of medications, and sleep, interact. The precise clinical and EEG phenotype of a single patient is often difficult to ascertain. Genetic studies have to rely on the ILAE clinical and EEG criteria.

If these ILAE criteria are helpful in the clinical and therapeutic management of IGEs, such is not the case in epidemiologic surveys. Many of these studies tend to confuse cryptogenic and idiopathic groups. For example, in rural Iceland, Olafsson and colleagues classified 62% of the idiopathic cases as being of unknown origin (2). Prior to the first ILAE classification, IGEs were classified as primary generalized epilepsies, and the classification included patients with various clinical seizures and the absence of brain lesion, regardless of the EEG features. It is evident that clinical criteria (e.g., seizure type, age of onset, personal and family background, evolution, response to drugs) and EEG criteria (e.g., bilateral spike-and-wave discharges) remain, at times, indispensable for diagnosis and inclusion in an epidemiologic study.

Address correspondence and reprint requests to Pierre Jallon at Epilepsy and EEG unit, CH 1211 Geneva 14, Switzerland. E-mail: pierre.jallon@hcuge.ch

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Some epileptic syndromes observed in infancy, such as benign familial neonatal seizures (BNFS) and benign myoclonic epilepsy in infancy (BMEI), should be considered as IGEs. These syndromes are quite rare and are usually excluded from epidemiologic surveys because they are difficult to observe and to classify.

Some time may elapse between onset of myoclonic seizures and the first medical visit. This time lag was estimated to be 1 year or longer in 37% of patients with absence seizures (3). In the CAROLE study, there were more patients with IGEs in the newly diagnosed epilepsy group than in the first-seizure group (33.8% vs. 17.4%) (4).

IGEs encompass several different syndromes, and a few patients shift from one phenotype to another. The diagnosis of juvenile myoclonic epilepsy (JME) is often made when a tonic-clonic seizure occurs. Recently, we observed a 24-year-old woman who had presented typical absences during infancy and myoclonic seizures during adolescence, and was currently hospitalized for absence status. When the status was resolved, we recorded on the EEG the typical pattern of phantom absences syndrome recently described by Panayiotopoulos et al. (5). How should this epilepsy be classified?

IGEs are usually easy to diagnose when clinical and EEG data are collected, but epilepsy is not synonymous with epileptic syndrome. The diagnosis of IGE may be made after an isolated seizure even if the traditional definition of epilepsy is that of a disease characterized by repeated seizures. Conversely, strict diagnostic criteria may lead to an underestimation of IGEs. EEG recordings cannot be performed in some population studies, especially in some developing countries. Furthermore, the main limitation encountered in the classification of idiopathic syndromes is the sensitivity of the initial EEG in detecting a diagnostic abnormality. When an EEG has been performed, it may remain inconclusive for the diagnosis, for example, in a patient with only tonic-clonic seizures. In a survey by Loiseau and colleagues in southwest France, 66 patients immediately fulfilled the diagnostic criteria

for IGE (6). Within 1 year, 13 patients previously classified as having cryptogenic or unclassified epilepsy or isolated seizures were reclassified as having IGE because bilateral spike-and-wave discharges were found on later EEGs. Recently, Marini et al. have shown the value of a directed EEG strategy; early postictal EEG followed by a sleep-deprivation EEG when the early EEG was negative led to a higher diagnostic yield in first-seizure cases in patients of all ages (7).

The case classification method is one of the pitfalls of epidemiologic studies in epilepsy and accounts for the discrepancies in the literature regarding incidence and prevalence rates. To illustrate this point, Loiseau et al. (8) attempted to classify 986 patients consecutively observed either in a university hospital (n = 344) or in private specialized practice (n = 642). IGEs were more numerous in the private practice sample (35.8 vs. 16.3) for two obvious reasons: IGEs have an age-related onset and are electroclinically easy to diagnose. The reported proportion of idiopathic cases in a study is quite different if the cohort is composed of children or adults, or both. Very few studies report incidence and prevalence rates; overall rates are difficult to calculate because only small numbers of cases are usually included.

Despite these remarks and restrictions, we can find some interesting data in the literature.

DISTRIBUTION OF IDIOPATHIC GENERALIZED EPILEPSY

Distribution of IGEs in some selected cohort studies

When we consider cohorts of adults and children, the general frequency of IGEs can be estimated at 15–20% (Table 1); however, there are two extremes. A study by Manford et al. found a frequency of 6.8%, while a study by Gastaut et al. found a frequency of 28.4% (9,10). In the CAROLE study, Jallon et al. reported a frequency of 16% in the single-seizure cohort and 27.4% in the index-seizure cohort (4). These rates are similar to those observed in rural Iceland (17%) and in a study by Granieri et al. in

TABLE 1. Frequency of generalized idiopathic epilepsies in some cohorts studies

Authors (yr)	Country	Population	Frequency (%)	I/P	Comments
Gastaut (1975)	France	4591 (C/A)	28.4		“Primary GE”
Joensen (1986)	Faroese islands	194	34.5	P: 2.9I: 14.8	“Primary GE”
Loiseau (1991)	France	1006 (C/A)	16.3 vs. 35.8		University hospital versus private practice
Manford (1992)	England	567 (C/A)	6.8		
Avanzini (1996)	Italy	8327 (C/A)	17.4 vs. 3.1		Definite cases versus uncertain
King (1998)	Australia	300 (C/A)	22.3		Consecutive cases
Zarelli (1999)	Rochester	157 (C/A)	15.7	I: 3.7	4 yr inclusion
Jallon (2000)	France	1942 (C/A)	16 vs. 27.4		Single seizure versus index seizures
Senanayake (1995)	Sri Lanka	1250 (C/A)	17.2		
Murthy (1998)	South India	2531 (C/A)	6.4		
Danesi (1985)	Nigeria	945 (C/A)	20.4		

C/A, children and adults; I, incidence; P, prevalence.

TABLE 2. Frequency of generalized idiopathic epilepsies in children cohort studies

Authors (yr)	Country	Population	Distribution (%)	Incidence
Oka (1995)	Japan	1872	20.6	I = 0.65
Sidenvall (1993)	Sweden	155	26.5	
Berg (1999)	US	613	20.6	
Endziniene (1997)	Lithuania	378	18.8	
Eriksson (1997)	Finland	157	23	
Waalder (2000)	Norway	198	12.1	
Sillanpaa (2001)	Norway	157	22	
Shah (1992)	India	470	15.3	
Kwong (2001)	China	309	32	

Lombardy (20.4%) (2,11). IGEs represented 25% of the total in 220 cases collected in the Department de l'Oise in France (12), which is in the same range as the 28% and 30% reported, respectively, by Cavazutti in school-aged children (13) and Cornaggia et al. in Italian army draftees (14). In developing countries where EEG is often not available, two studies (15,16) have reported similar distributions, but Murthy and colleagues reported a frequency of only 6.5% in India (17). In children's cohorts, the frequency is around 20% (18; Table 2). The incidence of IGE has been estimated in Rochester, Minnesota, USA, by Zarrelli et al. based on data from a small cohort of 157 patients collected from 1980 through 1984; the incidence rate was 3.7 per 100,000 persons (19). The same rate has been reported in Geneva (20); however, the rate was much higher in a study Joensen in the Faroes (2.9 per 1,000 persons) (21) and in Loiseau et al. study in southwest France (9.2 per 100,000 in persons less than 60 years of age and 18.4 per 100,000 for those under 25 years of age) (6). The prevalence of IGE was assessed to be only 0.65 per 1,000 persons in Lithuania (22).

Distribution of IGE syndromes

Benign familial neonatal seizures and benign myoclonic epilepsy in infancy

BNFS and BMEI are more recently defined, individualized syndromes, and very few cohorts included these syndromes in the distribution studies of IGE. BNFS are usually not included in epidemiologic studies. BMEI represents less than 1% of all epilepsies in children (4,6,23) and only 0.2% in a cohort of newly diagnosed epilepsies in children (18).

Absence epilepsies

Absence epilepsies are clearly age-dependent syndromes, and their distribution will vary if we consider children-only cohorts or combined adult and child cohorts.

In large cohorts, the frequency of childhood absence epilepsy (CAE) varies from 1.5% (24) to 12.1% (18). The variation depends largely on the mode and source of case definition. Gastaut et al. found 9.9% of patients

with absence epilepsy, but 17.8% were observed in populations under 15 years of age and only 2.8% in populations above 15 years of age (10). Similar rates were reported by Hauser: 6% in the total population, 12.8% in patients under 15 years of age, and 5.7% in patients older than 15 years (25). In developing countries, the rates are lower; Murthy et al. reported a rate of 0.5% (17), while Shah et al. reported 1.6% (26).

The incidence of CAE has been estimated to range from 0.7 per 100,000 persons (21) to 8 per 100,000 persons (27). In children's cohorts, incidence rates were variously reported as 7.1 per 6.3 per 100,000 persons, and 5.8 per 100,000 persons (3,6,28).

The cumulative incidence was 98 per 100,000 persons in a Danish study, and 98 per 100,000 in a Swedish report (3,29).

The prevalence of CAE has been found to range from 0.1 per 1,000 persons (28) to 0.7 per 1,000 persons (11). In the Lombardy study by Granieri et al. (11), the incidence was 18.8 in patients aged 0–9 years, 12 in patients aged 10–19 years, and 2.2 in patients aged 20–39 years. No cases were found in persons aged 40 years or older. In children's cohorts, the prevalence has been estimated to range from 0.4 per 1,000 persons (30) to 0.7 per 1,000 persons (31). Very few cases were reported in older persons; most of the time, they were reported as absence status epilepticus (32).

With some exceptions, a two- to fivefold preponderance of CAE in girls is usually reported (11.4% vs. 2.5%) (33).

Juvenile absence epilepsy

The epidemiology of juvenile absence epilepsy (JAE) has not been well studied because it is underdiagnosed; in many patients, only tonic-clonic seizures are recognized and the absences are overlooked. JAE seems to be less frequent, representing 0.2–2.4% of some large cohorts (4,24,34). The prevalence was estimated at 0.1 per 1,000 persons (28). In one study, JAE was as frequent as JME (35). This syndrome would represent about 20% of IGEs (2,36).

Juvenile myoclonic epilepsy

The exact frequency of JME is often difficult to assess, because this diagnosis is often made late and retrospectively confirmed when a tonic-clonic seizure occurs. The incidence of JME has been estimated at around 1 per 100,000 persons (6,21,28,37). The prevalence ranges from 0.1 per 1,000 persons (28) to 0.2 per 1,000 persons (21). The frequency in large cohorts is estimated to range from 5% to 10%. Other estimates of frequency include 2% by Berg et al. (34), 3.5% by the OREp group (23), 4.1% by Gastaut et al. (10), 5.3% by Jallon et al. (4), 5.4% by Tsuboi (38), 5.1% by Waalder (33), 7–9% by Janz (39), and 11.4% by Wolf and Goosses (40). In developing countries, Oka et al. reported no cases of JME (24); it represents only 0.98% of cases in a study by Danesi (15), 5% of cases in India

as reported by Murthy et al. (17), and 10.7% in a study conducted in Saudi Arabia by Obeid and Panayiotopoulos (41). Surprisingly, Sennayake and Roman report JME in 9.2% of their cohort (representing 53.7% of IGEs) (16) as opposed to a more realistic figure of JME representing 17–18% of all IGEs (16).

Epilepsy with grand mal on awakening

Generalized tonic–clonic seizures (GTCS) or grand mal seizures occur in a wide variety of epilepsy syndromes and can be attributed to multiple etiologies. This diagnosis can be difficult to make, because the patients also have minor seizures such as absences and myoclonias. When a patient has only GTCS and meets the criteria for IGE, it would be more appropriate to classify the condition as IGE with GTCS. There are few reliable epidemiologic data on epilepsy with grand mal on awakening (EGMA). Wolf (42) in a review of five articles on the chronology of grand mal seizures, found that the rates of EGMA varied from 22% to 37%. GTCS, without any precision on the conditions of occurrence, represent 23% of patients in the Rochester study, and no cases of EGMA are reported in some studies (9,19,21,33). In the southwest France survey, the incidence of EGMA was 1.8 per 100,000 persons (6,34).

Epilepsies provoked by specific modes of activation

This classification essentially includes photosensitive epilepsies. Photosensitive epilepsies represent 0.5% (34), 0.7% (23), 1.08% (4), and 1.5% (6) of epilepsies, and 2.3–6.7% (4,34) of IGE syndromes. Shah et al., in their Indian cohort, found that photosensitive epilepsies represent 7.9% of total epilepsies and nearly 35% of IGEs (26). The incidence of photosensitive epilepsies is 1.5 per 100,000 persons (6).

Other idiopathic generalized epilepsies

When a typical syndrome, as we described before, is not clearly individualized, the seizures presented by the patients are referred to as other IGEs. They represent 3.2% (10), 3.9% (6), 6% (23) and 9.7% (4) of large cohorts and 30% (23), 44% (4), and 55.6% (33) of the IGEs.

Absence status epilepticus

Absence status epilepticus (SE) can occur in patients with absence epilepsy at any time of life. In the majority of the cases, however, absence SE occurs before the age of 20 years. The occurrence of absence SE can be the first manifestation of IGE. Another subgroup of patients who may experience absence SE is made up of older patients with no history of seizures, whose seizures are provoked by excessive use of psychotropic drugs, withdrawal from benzodiazepines, or other toxic-metabolic causes. In the French-speaking Switzerland study (EPIS-TAR), which looked at the incidence of SE in a general population, Coeytaux et al. included four cases of absence SE, which represented 2.3% of the total SE cases (incidence rate of 0.6 per 100,000 persons) (32). However, two

cases were observed in patients with absence epilepsy and two were absence status de novo in older patients that were provoked by benzodiazepine withdrawal. In the EPISTEF study, Latour et al. observed 2.2% of patients with ASE (43). All the patients had a history of absence seizures. Nearly 3% of patients with absence seizures could present an absence status, and nearly 10% of adults who continued to have absence seizures that began in childhood were at risk for an episode of absence SE (44,45). Myoclonic SE in relation with JME seems very scarce, and no epidemiologic data are reliable.

MORTALITY

Mortality and the increased risk of mortality are evaluated by the standardized mortality ratio (SMR), which is the ratio of observed deaths to expected deaths in a population of patients with epilepsy, based on the age- and sex-specific mortality rates in a reference population. SMR is usually more elevated in patients with symptomatic epilepsy (46–48) than in patients with idiopathic epilepsy. In most studies of mortality, the term idiopathic epilepsy is often used in a broader sense and includes cryptogenic cases, or is grouped within a larger term such as epilepsies of unknown etiology. When the term idiopathic is used following the restrictive ILAE criteria, the mortality data concerning patients with idiopathic epilepsies do not show an increased SMR (49–52). Nevertheless in two studies, the SMR of patients with all types of idiopathic epilepsies was raised to 1.6 (1.0–2.4) (47) and 1.8 (1.4–2.3) (46). In the U.K. cohort, SMR for idiopathic epilepsies was no longer significantly increased (1.3; 0.9–1.9) after a longer follow-up (47).

CONCLUSION

IGEs are represented by well-known, individualized syndromes. Nevertheless, the evaluation of the frequency of some syndromes remains difficult, because the inclusion criteria are not respected, the EEG is not available, or the classification of the syndrome is not so easy. It is important to understand the frequency of these syndromes better, especially their worldwide distribution, because future genetic studies of epilepsy need to rely on the electro-clinical criteria.

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