Relevance of a family history of seizures

Few types of epilepsy are inherited as single gene disorders and, in general, this is the case for conditions that are common. However, most types of epilepsy have a weak genetic component and the presence of a family history increases the risk of an affected child. This applies only to those who are closely related to the affected member—that is a first degree relative—such as siblings, parents, or offspring. If the person is more distantly related and the family history concerns aunts, uncles, or grandparents, the risks are not very different from those faced by the general population.

Empiric risks are available for some of the more common types of epilepsy. The calculations of risk depend on an accurate diagnosis of the type of seizure and imply that sufficient investigations have excluded secondary causes of epilepsy.

Grand mal seizures

(1) In one close relative. If the frequency of any type of seizure (including isolated convulsions) is sought in the siblings of children of the proband, 11% of siblings and 14.3% of offspring will be affected. The background against which such a risk should be measured is a cumulative one, up to 20 years of age, of 4.1% for any type of seizure and 1.1% for epilepsy.¹ If a parent is affected the risk for a child with epilepsy is about 1 in 25 (4%). There is no increased risk of febrile convulsions in the offspring. If it is known that a parent has 3 cycles/second spike and wave activity, the 1 in 25 risk should be increased to 1 in 12.²

The risks to nieces and nephews are no greater than for the general population. Despite conflicting evidence most geneticists would add to the general risk a further risk for a child with a congenital malformation if the mother is affected and, because of the severity of the seizures, had remained on anticonvulsants during the pregnancy. Such a risk would be 6% compared with 2% for the general population.³

(2) In more than one close relative. If a parent (say a father) and his brother have had recurrent seizures the risk should be increased on an empiric basis to 10%. If more members are affected then single figure risks are appropriate.

(3) In a child with normal parents. The risk to these parents for another child with the same problem is about 1 in 25.

Petit mal

If there is a family history of petit mal the risks are not significantly different from grand mal (that is 1 in 25) for siblings or offspring. If there is electroencephalographic evidence of 3 counts/second spike and wave activity then the risk is double—that is 7–8%. Petit mal when present in families seldom breeds true and the increased risk is for either grand mal or petit mal. These risks are reasonable ones to take and if only one close family member is affected, they should be acceptable to most people.

Febrile convulsions in a previous child

One in 33 of children in the general population will have a febrile convulsion. The risk after one affected child is about 20% (1 in 5). If both parents and a previous child had febrile convulsions, the risk for another sibling increases to 1 in 3. These are clearly high risks but if on average only 1 in 10 proceed to recurrent non febrile seizures, the burden is not sufficient to deter most people. Few people seek advice but if they did most counsellors would be cautiously optimistic.

Infantile spasms

The most common circumstance is the family with a child who suffers from infantile spasms. As many of these children have severe developmental delay the burden of the disorder can be considerable. Tuberous sclerosis contributes about 10–20% of such patients. If this is excluded, and it is current practice to have the child looked at under a Wood's lamp, the recurrence risk is small. Fleischer *et al.*⁴ calculated a risk of 1.5%. If the affected relative is more distant than a first degree relative, the risk would decline to that for the general population.

Lennox-Gastaut syndrome

There is conflicting evidence about the role of genetic versus perinatal and postnatal factors. Some authors suggest that genetic factors probably play only a minor role but this has not been adequately explored. One study, Doose *et al.*⁵ suggests risks of 7%.

Benign epilepsy with rolandic spikes

The seizures are usually self limiting and a family history is present in about 15% of all patients. If the

family history concerns a previously affected child, recurrence risks for seizures may be as high as 30%. Even a risk of this magnitude may be acceptable for a self-limiting condition.

Focal epilepsy

There is an increased risk to offspring even if the parents have focal seizures, but such a risk is only 1–2%. Most of the risk is for generalised epilepsy rather than focal seizures and few people would be deterred by a risk of this order.

Benign convulsions of childhood

Several families have been recorded in which, despite an extensive family history, the affected child has seizures within the first few days of life which continue despite medication and then stop spontaneously. The pattern of inheritance is dominant and there are no long term sequelae. The family history will be clear and indeed the diagnosis should not be made without it.

Summary

The approximately tenfold increase in risk (1 in 250 to 1 in 25) to those who have a positive but not extensive family history of recurrent seizures would seem to be considerable but the actual figure of 4%

is small. Only if the family history concerns at least 2 closely related members does the risk reach the 10% mark (on the borderline between high and low), but even then the burden of the disorder and response to treatment in the other family members should be taken into account. Familial epilepsy often responds more readily to therapy than other types.

References

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M BARAITSER
The Hospital for Sick Children,
Great Ormond Street,
London WC1N 3JH