

Annotations

Nosology of febrile convulsions

Febrile convulsions have been defined as follows: 'An event in infancy or childhood, usually recurring between 3 months and 5 years of age, associated with fever but without evidence of intracranial infection or defined cause. Seizures with fever in children who have suffered a previous non-febrile seizure are excluded.'¹

It is common practice to divide convulsions with fever into benign and complex varieties,² and some would even exclude the latter from any discussion of febrile convulsions.³ This view was emphasised in a series of publications in the 1950s,⁴⁻⁶ especially those of Livingston.^{6,7} He defined two groups of patients on the basis of the duration and characteristics of the convulsions. Brief generalised convulsions constituted in his view 'simple febrile convulsions', and the risk of epilepsy was 3%. Prolonged or focal convulsions were an indicator of 'epileptic convulsions precipitated by fever', with a 97% risk of epilepsy. No subsequent authors have been able to define a group of children presenting with febrile convulsions who have such a high risk of developing epilepsy. The largest study of febrile convulsions yet performed (the National Collaborative Perinatal Project—the NCPP study) defined three factors associated with an increased risk of epilepsy by the age of 7 years.^{8,9} These were epilepsy in a parent or sibling, neurological or developmental abnormality noted before the first convulsion, and a complex convulsion—that is, a convulsion lasting more than 15 minutes, focal features, or more than one convulsion in 24 hours. Epilepsy occurred by the age of 7 in 2% of children with one of these risk factors and 10% of those with two. In defining complex convulsions duration is probably more important than focal features. Follow up data on brief focal convulsions are lacking, but three acknowledged experts in the field of childhood epilepsy recently agreed that such convulsions share the benign prognosis of brief generalised convulsions.¹⁰

Are prolonged febrile convulsions essentially different from brief ones? In a study of status epilepticus in children published in 1970 Aicardi and Chevrie concluded that prolonged and brief febrile convulsions differed 'in severity, not in nature'.¹¹ In the NCPP study prolonged convulsions (>15 minutes) led to epilepsy in 1.4% of neurologically normal children and in 9% of neurologically abnor-

mal children.⁹ Seventy four children had a first febrile convulsion lasting for 30 minutes or more, and three of them developed epilepsy, a risk not significantly greater than that of children with brief first convulsions. A British study of 13 135 children followed from birth to 5 years of age failed to show any intellectual deficit in children who had had febrile convulsions, either simple or complex (complex being defined as in the NCPP study).¹² The evidence suggests that prolonged febrile convulsions in neurologically normal children are not essentially different from brief ones, but in neurologically abnormal children the problem is a different one. Aicardi has suggested that prolonged febrile convulsions may be of two types, possibly equal in numbers, some resulting from pre- or early post-natal brain insult and the rest presumably of genetic origin.¹³ Adverse perinatal factors did not correlate with later epilepsy in the NCPP study, though there was such a correlation in Wallace's much smaller study in which 70% of the children were neurologically abnormal.¹⁴

Discussion and study of the problem of febrile convulsions is best served by adopting an inclusive definition within which subgroups may be delineated. Children who are neurologically abnormal are an obvious subgroup and their prognosis seems more likely to be related to their neurological abnormality than to their febrile convulsions. Further studies are needed in which specific types and degrees of neurological abnormality are related to outcome. To refer to unspecified 'neurological abnormality' as a study factor is a gross and confusing simplification dictated by the tyranny of study numbers. In the NCPP study 34 children developed epilepsy by the age of 7. Fourteen of the 34 were mentally retarded, and seven of the 14 had cerebral palsy. None of the 14 children were known to have been normal before the first febrile convulsion.⁹ A retrospective survey of 40 years' experience in Rochester, Minnesota, showed a very high incidence of epilepsy (40%) after febrile convulsions in children with mental retardation and cerebral palsy.¹⁵ In Wallace's study of 112 children in Edinburgh 79 had chronic neurological disorder, and, of those, 12 had epilepsy eight to 10 years later.¹⁴ In the British cohort study only children with neurological abnormality preceding the febrile con-

vulsions had an adverse outcome as regards intellectual function at the age of 5.¹²

A first degree family history of epilepsy increased the risk of epilepsy after a febrile convulsion in neurologically normal children from 1% to 5%.⁹ This is little greater than the risk of epilepsy with the same family history irrespective of febrile convulsions (2–5%).¹⁶ Such a family history was found in 5.6% of children in the NCPP study,⁹ in 1.8% in the Rochester study,¹⁵ and about 2% in a Danish study.¹⁷

In the NCPP study a complex first convulsion in a neurologically normal child was associated with little increased risk of epilepsy (1.7% instead of 1.1% after an uncomplicated convulsion), but in a neurologically abnormal child the risk rose to 10%.⁹

In the Rochester study with much longer follow up epilepsy followed focal or repeated convulsions in 7% of neurologically normal children and 40% of neurologically abnormal children.¹⁵ Convulsions lasting for 10 minutes or more were followed by epilepsy in 10% of the whole series, in 6% of those who were neurologically normal and whose convulsions were generalised and single, and in 20% of those who were neurologically abnormal or whose convulsions were focal or repeated in the same episode.

The age of the child at the time of the first febrile convulsion seems to be important. The risk is higher if the first convulsion occurs in infancy, especially in the first 6 months,^{9 15 18} but to what extent this is simply a reflection of the younger children being more likely to be neurologically abnormal is unclear. In the Rochester study children aged between 2 and 4 years at the time of the first convulsion had no increased risk of epilepsy.¹⁵

The main factor associated with recurrence of febrile convulsions is the age of the child, younger children being more prone to recurrence.^{9 19 20} There are conflicting data on whether or not complex convulsions are more likely to recur. The NCPP study, that of van den Berg in California,¹⁹ and the British cohort study¹² did not show any correlation between the characteristics of the first febrile convulsion and the risk of recurrence, but a recent study from Denmark found the following factors to be related to the risk of recurrence; age 15 months or less, complex first convulsion, family history of epilepsy or of febrile convulsions, and day nursery care.²⁰

How, in practice, should a paediatrician assess prognosis in a child who has had a febrile convulsion? By far the most important factor is the child's neurological and developmental state. A neurologically normal child has a low risk of epilepsy and virtually no risk of other neurological or develop-

mental handicap. When neurological abnormality is discovered the prognosis will depend on the degree and type of abnormality, and if the abnormality is detected along with convulsions in early infancy the prognosis will be correspondingly poorer.

Status epilepticus is rightly feared by paediatricians and should be avoided by early and effective treatment of convulsions. In the series of 239 cases reported by Aicardi and Chevrie signs of severe neurological damage followed the episode in a high proportion of cases.¹¹ Series of patients from specialised paediatric neurology referral centres, however, often give a more gloomy impression than community based series, and of the 1706 children in the NCPP study 74 (4.3%) had a first febrile convulsion lasting for 30 minutes or more, and none had a persisting neurological defect resulting from the seizure. Seven of the 1706 children had a Todd's paresis, and one developed focal epilepsy affecting the same side as the Todd's paresis.

The following practical division of febrile convulsions is proposed:

- (1) Neurologically normal children—good prognosis, probably 1–5% risk of epilepsy depending on adverse features of convulsion.
- (2) Neurologically abnormal children—prognosis depends on degree and type of abnormality and may be particularly poor in very young children.
- (3) First degree family history of epilepsy—associated with an increased risk of epilepsy (2–5%) irrespective of febrile convulsions.

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