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SUPPORTING EPILEPSY STUDIES

Understanding Panayiotopoulos Syndrome

Colin Ferrie

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ABOUT THE AUTHORS

Colin Ferrie

Colin Ferrie is a consultant paediatric neurologist in Leeds. His subspecialty interest is in the epilepsies. His service provides comprehensive services for children with epilepsy in Leeds and the Yorkshire region.

He trained in Manchester, Edinburgh and London, being an epilepsy research fellow at Guy's and St.Thomas's Hospitals between 1992 and 1995. He obtained an MD for his studies into the use of PET scanning in the investigation of childhood epileptic encephalopathies. Other research interest have included the idiopathic focal epilepsies of childhood (esp. Panayiotopulos syndrome), absence epilepsies and febrile seizures.

He is on the executive committee of the British Paediatric Neurology Association, the board of the European Paediatric Neurology Society and has served on the Council of the UK Chapter of the International League Against Epilepsy.

AN Other

AN Other is the parent...

1. WHAT IS PANAYIOTOPOULOS SYNDROME?

Epilepsy

Epilepsy is not a single disease but rather a large group of disorders whose common feature is the tendency to have recurrent epileptic seizures. Epileptic seizures are events which are caused by abnormal electrical discharges in the brain. They can take many different forms, ranging from brief, blank stares to convulsions (characterised by body stiffening and repetitive limb jerking). Because, epilepsy is the tendency to have recurrent epileptic seizures, it is usually not formally diagnosed until at least two have occurred. Many different underlying problems can give rise to epilepsy. It can be a consequence of another underlying problem, such as cerebral palsy, a brain tumour or a head injury. This is sometimes called symptomatic epilepsy. If no cause for the epilepsy can be found, and there is nothing to suggest any other problem with brain functioning (such as learning difficulties), the epilepsy is called idiopathic epilepsy. In recent years it has become clear that many idiopathic epilepsies are a consequence of abnormalities or variations in our genetic make-up. Such epilepsies are now often called genetic epilepsies. It is important to note that just because a disorder is genetic in origin it need not be inherited. Therefore, although some people with genetic epilepsy have close relatives who also have epilepsy, many do not. If no underlying cause can be found for the epilepsy but other problems, such as learning difficulties, point towards an underlying brain problem, the epilepsy can be called cryptogenic (meaning hidden) or probably symptomatic.

Epilepsy in Children

Some types of epilepsy occur in all age groups but others only or mainly occur in particular age groups. There are many epilepsies, which begin in children and usually end before adult life. These can be called the childhood epilepsies. Childhood epilepsies mostly seem to be genetic epilepsies. One reason why some genetic epilepsies only occur in children is that some genes are only active (the technical term is 'expressed') during certain periods of development. As such, children 'grow out' of these epilepsies as they develop neurologically.

Some epileptic seizures involve widespread parts of the brain (including both sides) from the start of the seizure. These are known as generalised seizures. Others begin and may remain confined to a localised area in the brain. These are known as focal epileptic seizures (the terms partial and localization-related may also be used and mean exactly the same). Some focal seizures spread to include both sides of the brain. These can be called secondarily generalised (focal) epileptic seizures. PS is idiopathic epilepsy that occurs in childhood and the seizures that characterise it are focal. In some children with PS secondarily generalised seizures can occur. As with other childhood epilepsies, PS may be genetic in origin but this has not yet been proven.

Seizures in Panayiotopoulos Syndrome (PS)

In PS the seizures are characterised by disturbances in the autonomic nervous system (for this reason they are often classified as focal autonomic seizures). The autonomic nervous system is responsible for controlling functions without the need for conscious control, for example our heart rate, breathing pattern, skin blood flow, temperature and the dilation/constriction of our pupils. Vomiting, and the retching and nausea which may accompany it, are also controlled by the autonomic nervous system.

During seizures in PS many different autonomic nervous system disturbances have been described but most often they begin, often rather inconspicuously, with a change in the child's behaviour (e.g. irritability) and with the child complaining of feeling sick. Sometimes retching and/or vomiting begin out of the blue. The child may look pale or flushed, their breathing may become irregular or shallow and their lips may become blue. Sometimes coughing or salivation is prominent and the pupils are often markedly larger than usual (dilated). Less commonly they become very small, like pin-points (constricted). The child's temperature may be increased, or the child may complain of feeling cold or hot when their temperature is normal. Some children may be incontinent of urine or faeces.

At the start of the seizure children are usually fully conscious and able to respond to questions. However, as the seizure continues consciousness usually becomes impaired and the child may become increasingly confused, may appear disoriented or distant and will

no longer respond appropriately. With this impairment of consciousness it is common for the eyes and/or the head to move to one side and remain fixed in this position (this is known as aversion). By now, and depending on the circumstances of the seizure, the child is likely to be lying down and most of their body will be floppy.

Many seizures in PS end without there being any convulsive movements (that is rigidity of the body with repetitive jerking of the limbs). However, some terminate with convulsive movements, which may appear as small as twitches, and may be down one side of the body (a hemiconvulsion) or affect the whole body (a convulsion). In PS seizures can be short, but more often last many minutes and sometimes even hours. Seizures lasting over 30 minutes are called 'status epilepticus'. PS is one of the common causes of non-convulsive status epilepticus. Afterwards the child is likely to be tired and may be confused or moody for some time. They may go into a deep sleep. Once rested the child will recover completely.

It is not known why seizures in PS occur, and they may appear to 'come out of the blue'. However a majority occur whilst the child is sleeping (the child often rouses briefly form sleep immediately before the seizure), including day-time naps. Some parents notice that seizures are more likely to occur under specific circumstances, such as if the child is tired or hungry or if he or she has been emotionally upset.

2. DIAGNOSIS & ASSESSMENT

Symptoms

The characteristics of PS seizures are for the most part not typical of those expected during epileptic seizures. For this reason epilepsy is often not suspected, especially with initial seizures. Because relatively few children are diagnosed with PS, many general and emergency medical practitioners are unaware of PS and the symptoms of its seizures. The rate of misdiagnosis is high. Children with PS are sometimes thought to have car sickness (because the seizures often occur during daytime naps in the car), abdominal migraine or cyclical vomiting (because of episodic episodes of vomiting), or faints (because of the floppiness and pallor often seen). In addition some children are admitted to intensive care units during prolonged seizures and may be suspected of having suffered a severe brain insult. Their rapid recovery to complete normality may come as a shock- albeit a very welcome one!

PS has been described in children of all ages but most characteristically starts around the ages of 4-6 years. It usually occurs in children who have no other problems and who are developing normally. However, there is often a history of previous febrile seizures. Many children suffer only a single seizure and in most cases even if seizures are recurrent, the total seizure count is low (e.g. a handful). There are, however, children who have frequent seizures of PS (meaning they may over a period of months or years have say a dozen or more seizures). Seizures are generally only active for a few years and children with PS are expected to 'grow out of their seizures' before (usually well before) they reach 16 years of age.

Brain Scans And EEG

The diagnosis of PS is mainly made on the basis of a good description given by someone who has witnessed one or more of the seizures. Because the seizures are often prolonged, many parents are able to record them on their mobile phones. This can be extremely helpful. There is no single test which can diagnose PS. Brain scans (CT and MRI) are expected to be normal. However, an EEG can be helpful. An EEG (which stands for electroencephalogram) is an investigation in which the electrical activity of the brain is recorded using a large number of electrodes applied to the scalp. In the past the 'brain

waves' were recorded onto paper. Now they are recorded digitally and displayed on a screen. Most EEGs are recorded when awake for between 20 and 40 minutes (a standard EEG). The EEG can also be recorded during sleep (a sleep EEG). There are also methods for recording EEGs over prolonged periods of time (days). Recording an EEG is entirely safe. It does not hurt and can even be fun for the child.

Some children with PS have persistently normal EEGs. However, most show features, known as epileptiform abnormalities. These are brain waves not usually seen in people who do not have epilepsy but which are often seen in those who do. Examples of epileptiform abnormalities which might be encountered in PS are 'posterior spike-wave discharges', 'occipital paroxysms' and 'cloned like-repetitive complexes'. These are illustrated in the picture below. None are exclusive to PS in that they can occur in other forms of epilepsy as well. However, the combination of a description of symptoms that fits PS with an EEG showing such features may allow a confident diagnosis to be made. Sleep activates the EEG features of PS. Therefore, if an awake EEG is normal, obtaining a recording during sleep can be helpful.

3. TREATMENT & OUTLOOK

Drugs Or No Drugs

The seizures of PS generally respond well to conventional antiepileptic drugs (commonly used drugs include carbamazepine, sodium valproate and lamotrigine). However, given the low seizure count the fact that seizures are only active for a few years, and the potential side effects of conventional antiepileptic drugs, many children are managed without regular antiepileptic drug treatment. If prolonged seizures are a problem so-called rescue treatment (given during the actual seizure) with either rectal diazepam or oral midazolam can be used. These drugs act to reduce the over excited brain activity that happens during a seizure and thus terminate the seizure. Sometimes parents administer this rescue medication at home without the need for medics to be present or for subsequent hospitalisation. Although it is rare, some children with autonomic seizures such as those in PS can be sensitive to these drugs, in which case reduced dosage can be helpful and hospital admission or paramedic presence may be advisable before administering the rescue medication. It can be difficult to determine that a seizure has definitely terminated after administration of rescue medication. Good indicators are a normal breathing pattern, a more neutral pallor (neither pale nor flushed) and the pupils in the eyes responding to light.

The autonomic disturbances which characterise seizures of PS have caused concern that seizures in PS might be associated with cardiac rhythm disturbances or cessation of breathing leading to cardio-respiratory arrest. There are a few reports of children with PS receiving resuscitation during seizures. It is not possible to offer complete reassurance. However, it is clear that if seizures of PS pose any danger, the danger is exceedingly small.

Outlook

PS is often called a benign epilepsy. This roughly means 'not serious'.

Obviously the seizures themselves can be very frightening. However, there are no reports of any child with PS suffering any permanent neurological damage as a consequence of a seizure. Children with PS are expected to develop normally, although there are no detailed studies on this, so the possibility of associated learning or behaviour problems has not yet been excluded. PS does not continue indefinitely. Children grow out of it, usually within a year or so of it starting. However, some children continue to have seizures of PS during childhood and early adolescence.

A small number of children with PS develop other types of childhood epilepsy, particularly rolandic epilepsy, but children with PS are no more likely than any other child to develop epilepsy in adult life. While PS can be challenging and upsetting for children and those who care for them, but the prognosis for those with seizures of this nature is very good.

Disclaimer statement

The information and medical advice expressed here are not a substitute for talking to your doctor. This information should not be used in place of a visit, call or advice of a physician or other health care provider. The advice is intended to provide a general guide for a person to discuss their personal medical condition with their health care provider. You should not disregard medical advice or delay seeking it because of something you have read here.

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