UNDERSTANDING
PANAYIOTOPOULOS SYNDROME

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WHAT IS PANAYIOTOPOULOS SYNDROME?

Epilepsy

Epilepsy is not a single disorder but a 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. About 70% of epilepsies have a genetic basis.

Epilepsy in children

There are many epilepsies that begin in childhood and usually end before adult life. These are called the childhood epilepsies. Some epileptic seizures involve both sides of the brain. These are known as generalised seizures. Others begin in one side of the brain. These are known as focal epileptic seizures. Some focal seizures spread to include both sides of the brain. These can be called secondarily generalised epileptic seizures. Panayiotopoulos Syndrome (PS) is a focal epilepsy. In some children with PS secondarily generalised seizures can occur. PS is suspected to 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.

Many different autonomic nervous system disturbances have been described during seizures in PS. 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 or affect the whole body. In PS, seizures can be short, but more often last many minutes and sometimes even hours.

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 daytime naps. Some parents notice that seizures are more likely to occur under specific circumstances, such as if the child is tired or hungry or s/he is emotionally upset.

DIAGNOSIS & ASSESSMENT

Symptoms

The characteristics of PS seizures are quite unusual and not typical of other types of epilepsy. For this reason epilepsy is often not suspected, especially with the initial seizures. Because PS is rare, many clinicians in A&E or in general practice may be unfamiliar with PS and the symptoms of its seizures. Misdiagnosis is common. 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 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 PS seizure and in most cases even if seizures are recurrent, the total seizure count is low (less than five). There are, however, children who have frequent seizures of PS (up to a dozen over a period of years. Seizures are generally only active for a few years and children with PS are expected to 'grow out of their seizures' 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 from beginning to end. 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 number of electrodes applied to the scalp. 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). Recording an EEG is entirely safe, it does not hurt.

Some children with PS have persistently normal EEGs. However, most show features known as epileptiform abnormalities. Examples of epileptiform abnormalities which might be encountered in PS are 'posterior spike-wave discharges', 'occipital paroxysms' and 'cloned like repetitive complexes'. None are exclusive to PS. 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 an EEG recording during sleep can be helpful.

TREATMENT & OUTLOOK

Drugs Or No Drugs

The seizures of PS generally respond well to conventional antiseizure medications. However, given the low seizure count, the fact that seizures are only active for a few years, and the potential side effects of conventional antiseizure medications, many children are managed without regular drug treatment. If prolonged seizures are a problem then rescue treatment (given during the actual seizure) with buccal midazolam can be used. These drugs act to terminate the seizure. Often parents administer this rescue medication at home. 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 that characterise seizures of PS have caused concern that seizures in PS might be associated with cardiac rhythm disturbances or stopping 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, the outlook is very good.

Disclaimer statement

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