



Posterior fossa syndrome

Information for parents of a child with PFS

This factsheet has been written to help you understand posterior fossa syndrome (PFS), the name given to a collection of symptoms which may occur together following surgery to the back of the brain. PFS is also sometimes known as cerebellar mutism syndrome.

Symptoms are not always present immediately after surgery, but often develop in the following few days. Early symptoms include difficulties with speech, movement and swallowing. If your child develops PFS they may appear very different. It's important to remember they are still the same child on the inside.

The information in this factsheet explains what PFS is and its impact, and provides information about rehabilitation and recovery. If you have any questions, the medical team treating your child will be happy to answer them.

When does PFS occur and how long does it last?

PFS has been reported to occur in up to thirty percent of children undergoing surgery to the back of the brain. The severity of the condition varies considerably from one child to the next. It is therefore difficult to predict the extent or duration of their recovery.

However, the majority of children will make significant improvements. The extent of any long term difficulties may not be evident initially and become apparent later as the child returns to education and home life.

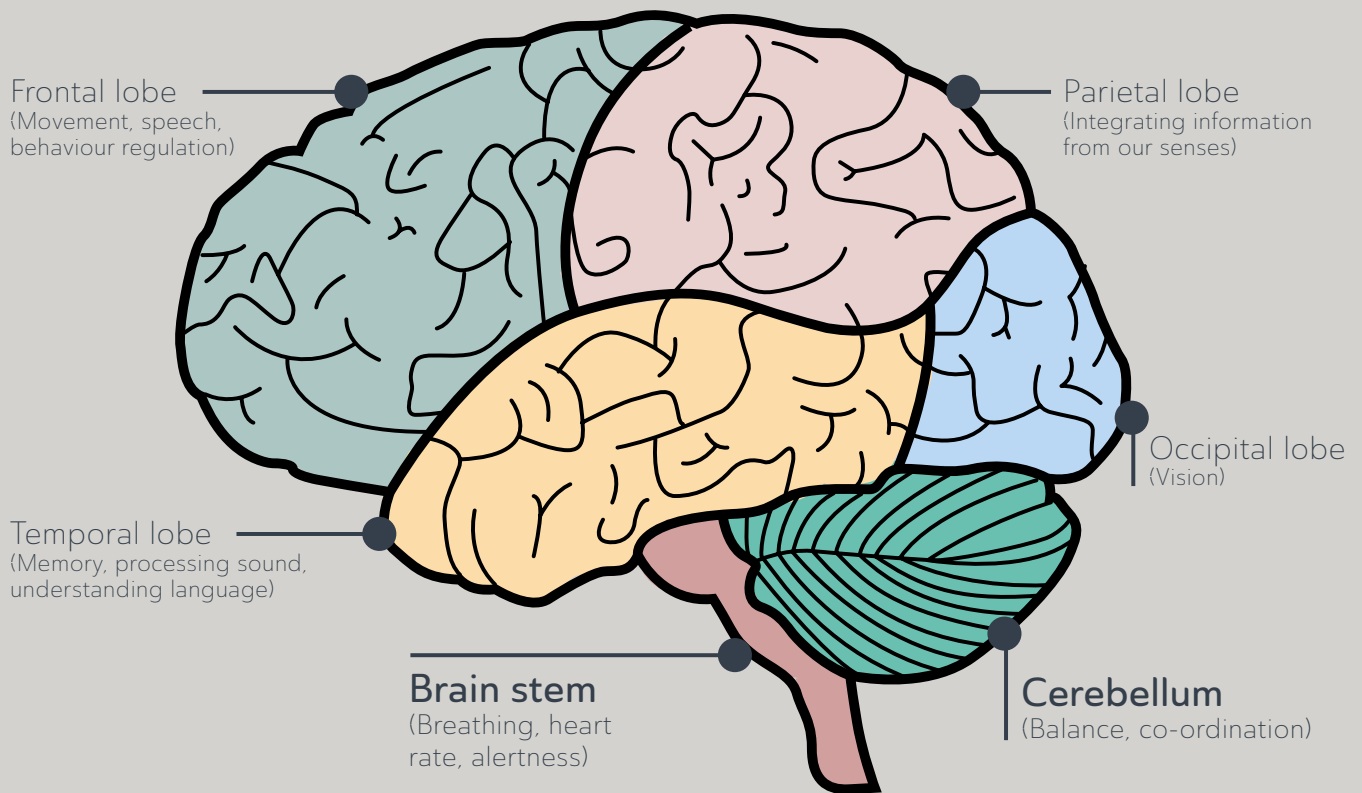
Ongoing rehabilitation and support may be required for months or even years.

Causes

The exact cause of PFS is unknown. It occurs following operations to the cerebellum (responsible for balance and coordination) and brainstem (responsible for basic functions such as breathing, heart rate and body temperature), particularly those to remove a type of tumour called medulloblastoma. Tumours that involve the brain stem and are located in the brain midline are also associated with PFS.

It is currently unclear why it affects some children and not others, and there is ongoing research into this subject. Current evidence suggests that PFS is caused by an interruption to a particular pathway in the brain that sends signals from the cerebellum to the parts of the brain that process information, sensations and body movements.

Areas of the brain and an approximate guide to their functions



PFS is identified by a collection of symptoms

Speech and communication

Reduced or absent speech is often one of the most striking features, which is why PFS is sometimes called 'cerebellar mutism'. Children may not speak at all or their speech may be slow or slurred and difficult to understand. This usually occurs within the first few days after surgery but may not be present immediately after surgery.

Body movements ('motor control')

Children can display low muscle tone (hypotonia), unsteadiness and problems with coordination (ataxia). They may not be able to coordinate voluntary movements, and can find it difficult to hold their head up or sit up.

PFS can affect the movements involved in chewing and swallowing. In severe cases, children may have difficulty swallowing their saliva, making it unsafe for them to have anything to eat and drink by mouth. Usually, children regain the voluntary movements of the lips and tongue first before being able to eat and drink, for example, being able to stick out their tongue and move it from

side to side. Children who have been severely affected may need tube feeding, especially when radiotherapy and chemotherapy are given at the same time.

Emotional difficulties ('emotional lability')

Many children and young people experience 'emotional lability', a term used to describe rapid changes of mood and can include sudden uncontrolled laughter or tears. They may also display angry behaviour such as shouting or swearing, which may seem out of character.

Mood swings and agitation can be very distressing for families to see and can be particularly evident after the initial stages of recovery following brain surgery. This can be made worse by a child feeling scared, or unable to communicate.

Learning and understanding ('cognition')

Due to the speech and movement problems seen in PFS, it can be difficult to assess the impact on children's learning abilities until they have made a significant recovery.

Nevertheless, it is important to consider that thinking and reasoning could be impaired and processing speed is likely to be slowed. This means that children may be confused, might find it difficult to make decisions, and can often take a long time to respond to questions.

The aim of rehabilitation is usually to recover skills, but sometimes other ongoing treatment can affect your child's functioning and therefore the aim at that point can be to help your child maintain skills.

A child may appear uncooperative and be reluctant or unwilling to undertake therapy during their rehabilitation and may need lots of patience and understanding.

“ If your child develops PFS they may appear very different. It's important to remember they're still the same child on the inside. ”



PFS symptoms will improve with time but recovery is different for each child.

Your child will need ongoing rehabilitation from a team of therapists to help their recovery in a range of different areas, depending on their individual difficulties.

See page 4 to read about the rehabilitation process.

Rehabilitation

Rehabilitation can take place in hospital, at home, and occasionally at a specialist rehabilitation centre depending upon your child's individual needs. Children who have more severe symptoms of PFS early on are more likely to have symptoms that last longer.

Your child's rehabilitation team will include the following specialists:

- **Speech and language therapists** can provide support for any speech, language and communication difficulties. In addition, they help with swallowing difficulties, supporting the process of eating and drinking and ensuring it is safe and pleasurable.
- **Physiotherapists** can provide assistance with maximising strength, balance and coordination of movements, helping children regain their physical skills.
- **Occupational therapists** support children to participate in everyday life activities, enabling children to regain their independence.
- **Clinical psychologists or neuropsychologists** can help to identify and support any difficulties with ongoing learning as well as supporting children and their families with emotional or behavioural issues as they adjust to what has happened to them.

Other therapists, for example, play therapists, art therapists and music therapists, may also be involved in your child's rehabilitation.





Recovery and long term prognosis

Motor control

For a few children it will take many months to regain their physical abilities. Most but not all will be able to walk independently. However, ongoing difficulties with balance, coordination and body movement are common.

Speech and communication

In most cases communication comes back very gradually with more spontaneous sounds first (laughing and crying) followed by individual sounds, slowly developing into single words and extending into short phrases. Some children may communicate non-verbally using head nods/shakes, gestures or communication boards initially.

Emotional difficulties

Emotional difficulties can persist after recovery has been made in many other areas. In addition to sudden emotional distress associated with emotional lability (often crying or appearing agitated), children are at

increased risk of emotional, behavioural and social problems following PFS. Children often feel frustrated if they are unable to participate in the activities that they used to enjoy. This may affect their confidence as well as social relationships. If you are concerned about ongoing low mood or behaviour problems do discuss this with a member of your child's rehabilitation team.

Learning and cognition

There are many factors that can affect learning and cognition in children with brain tumours, such as the tumour itself, surgery and radiotherapy. It is therefore difficult to establish how much of an impact PFS has on cognitive function in the long term. Research comparing children with the same type of tumour and treatment, with or without PFS, has found that children with PFS can have more difficulties in specific cognitive areas, such as attention and working memory. However, further research is needed in this area.

It's important to remember any learning difficulties will vary for each child. It is helpful for your child to have a neurocognitive assessment by a clinical psychologist, educational psychologist or clinical neuropsychologist to identify your child's particular strengths and weaknesses.

Returning home

Preparation for your child's discharge from hospital begins as soon as your child is admitted to hospital as it can take some time to arrange the right support.

The medical and nursing team will work closely with other professionals to assess your child's needs and any additional support required, both in the short and long term. This assessment should include consideration of medical needs, physical needs, psychological and practical needs and returning to school.

Medical needs

Information about any ongoing treatment for your child's tumour will be explained to you in detail, and you will receive information on how to care for your child at home. Referrals will be made to the appropriate community nursing team and other services you may need.

Physical needs

Your child will have an assessment to see how they will cope at home. If any equipment is needed, this will be ordered by the occupational therapy team. If required, information about more permanent adaptations to your home will be provided.

Psychological and practical needs

Members of the hospital team, including the outreach nurses and CLIC Sargent social worker, will be able to support you, and also advise on other support available locally.

Returning to school

Going back to school is an essential part of your child's ongoing rehabilitation. It is usually possible for your child to return to school on a reduced timetable and slowly build up the time they spend at school. Although your child may tire easily, being back at school will help your child get back into a routine and give them the opportunity to see their friends.

Make sure the school understands your child's needs so appropriate support is in place and your child can be included in normal school life as much as possible.

Additional support after leaving hospital

Whilst your child is attending hospital regularly it is likely that you will be able to access support from within the hospital. Upon discharge the hospital team should refer you to appropriate professionals in the

community for support. This can include community physiotherapy, occupational therapy and speech and language therapists.

If you feel that your child has needs which are not being met, discuss this with your specialist nurse or consultant who will be able to liaise with the relevant professional.

Leaving hospital can often be a difficult time for families who can feel that they are losing the safety net of the professionals who understand their child's unique needs. It may be useful to discuss these concerns with a member of the team, for example, your specialist nurse or CLIC Sargent social worker, who will be able to signpost you to support groups or other organisations who are able to help.

Getting support in education

Every mainstream school or nursery has a Special Educational Needs Coordinator (SENCo) who is responsible for ensuring that the needs of children with disabilities or special educational needs are met. Within colleges there is also a designated person for this role but they may not be called a SENCo.

In order to ensure that your child is supported effectively, it is helpful to discuss your child's needs with the school SENCo as early as possible. A member of the hospital team may be able to support you with this. It may also be possible to invite the SENCo or another senior member of school staff to the multidisciplinary team meeting (MDT) or discharge meeting so they can begin to understand your child's needs and ask questions.

An Education, Health and Care (EHC) plan looks at all of your child's needs across education, health and social care and ensures that the child receives the right amount of support by engaging everyone involved in the child's care. EHC plans are for all children with special education needs. An assessment will need to be requested by the school, hospital or parents and is carried out by the local authority.

Whether or not your child has an EHC plan, their progress and level of support should be regularly reviewed.



Further information about returning home after treatment for a brain tumour can be found in the CCLG booklet 'Brain and spinal tumours'.

CCLG also produces a booklet entitled 'Welcome back!', which offers advice and support for teachers helping children and young people return to school after a diagnosis of cancer.

“It's important you plan time for yourself and ensure you have the support you need.”

Supporting your child

Your child will need ongoing support and parents understandably find it very stressful to see their child upset. It is natural to try to find out what your child is upset about to make things easier for them. However, as biological changes in the brain can be associated with strong emotional reactions without any apparent trigger, your child may not know why they are upset. Whilst it is important to encourage your child to talk about their feelings, it is also important to know that at times, they will have no answer.

On those occasions it is helpful to have a plan in place of how your child can manage their distress until it passes. They may simply benefit from a hug, some quiet time, an object that is special to them, an activity that distracts them, or some rest.

Taking care of yourself

Seeing your child suffer from the symptoms of PFS can be very distressing. It is very normal to feel overwhelmed at times, especially as you already have to deal with your child's diagnosis and treatment. Being there for your child, providing reassurance and familiarity will help them begin their gradual process of recovery.

Whilst your natural instinct as a parent may be to give up everything to support your child, you also need to look after yourself to allow you to best support your child. Try to eat regularly (even if you don't feel like it) and rest when possible. The recovery process from PFS may take a long time. It is important that you plan time for yourself and ensure that you have the support you need in order to be able to be there for your child throughout their journey of recovery.

Sources of information and support

Useful organisations

Children's Cancer and Leukaemia Group

www.cclg.org.uk

CCLG is the UK and Ireland's professional association for those involved in the treatment and care of children with cancer, and is dedicated to improving best practice and outcomes. We support research and also provide accredited information for patients and families.

The Child Brain Injury Trust

www.childbraininjurytrust.org.uk

Helpline: 0303 303 2248

Provides information on children's acquired brain injury and offers training and support for professionals including school staff.

The Children's Trust Brain Injury Hub

www.braininjuryhub.co.uk

Provides information on acquired brain injury. Also provides a discussion forum for parents and families.

CLIC Sargent

www.clicsargent.org.uk

Helpline: 0300 330 0803

Provides practical support and advice for children and young people affected by cancer and their families and offers accredited information.

The Brain Tumour Charity

www.thebraintumourcharity.org

Provides information about brain tumours and their treatment.

The Danny Green Fund

www.thedannygreenfund.org.uk

A charity set up by the family of a child who experienced PFS. Provides information and funding for equipment and therapy.

Useful resources

There's a comprehensive range of free publications available for children and young people with cancer and their families from a number of Information Standard accredited organisations, including CCLG and CLIC Sargent.

Children's Cancer and Leukaemia Group (CCLG)

Children and young people with cancer:

A Parent's Guide (2014)

Welcome back! A guide for teachers helping children and young people returning to school after a diagnosis of cancer (2015)

Brain and spinal tumours (2016)

For schools

Cancer and school life pack (CLIC Sargent, 2014)

Welcome back! A guide for teachers helping children and young people returning to school after a diagnosis of cancer (CCLG, 2015)

Returning to school: A teacher's guide for pupils with brain tumours (Second edition, The Royal Marsden and Cerebra, 2015)



Children's
Cancer and
Leukaemia
Group

Children's Cancer and Leukaemia Group is a leading children's cancer charity and the UK and Ireland's professional association for those involved in the treatment and care of children with cancer. Each week in the UK and Ireland, more than 30 children are diagnosed. Two out of ten children will not survive their disease.

We bring together childhood cancer professionals to ensure all children receive the best possible treatment and care. We fund and support research into childhood cancers, and we help young patients and their families with our expert, high quality and award-winning information resources.

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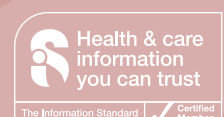
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For information about the sources used to put this publication together, please contact us.

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