



Great Ormond Street Hospital for Children NHS Foundation Trust

## Parry-Romberg syndrome: information for families

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**Parry-Romberg syndrome (also known as Progressive Hemifacial Atrophy) is a rare condition affecting the skin and soft tissues on one side of the face (hemifacial). It is considered to be within the group of conditions called morphoea and is named after the two doctors who first described it in the mid-19th century. This information sheet from Great Ormond Street Hospital (GOSH) explains the causes, symptoms and treatment of Parry-Romberg syndrome.**

The condition usually starts in childhood when the skin and soft tissues start to waste away (atrophy). The wasting usually continues for between two and ten years before becoming stable. The underlying muscle and bone may also waste away causing functional problems with eating and speech and an appearance difference. Once in its stable phase, it does not get any worse with age.

### What causes Parry-Romberg syndrome?

The cause of Parry-Romberg syndrome is not yet known but several theories have been put forward, including trauma, infections, hormonal problems and autoimmunity. It does not appear to be inherited as most cases happen sporadically (out of the blue). Further research is needed to work out what causes Parry-Romberg syndrome.

### What are the symptoms of Parry-Romberg syndrome?

The atrophy or wasting usually starts above the jaw or between the nose and upper lip. Over a period of months, it continues upwards to around the eye and ear and downwards towards the neck. The skin, soft tissue and muscle may

become wasted and start to develop a sunken appearance. As the tissue around the eye becomes affected, the eye can appear sunken.

The tissues inside the mouth – the tongue, gums and soft palate – also become wasted and tooth development may slow down or stop. This may cause problems with eating and speech. The bone in the skull and jaw may also be affected.

The skin is also affected, with areas with too much pigment (hyperpigmentation) which appear darker, as well as areas with too little pigment (hypopigmentation). Areas of facial hair, such as the eyebrows may lighten in colour and start to fall out (alopecia). There can be pain on the affected side (not common) and seizures or fits may develop.

Once the condition enters its stable phase, the wasting of skin, soft tissue and muscle stabilise but can be re-activated.

### How is Parry-Romberg syndrome diagnosed?

Doctors will take a clinical history of when and where the symptoms appeared and their severity, as well as a physical examination. Imaging scans,

such as x-ray, CT or MRI may be suggested to monitor bone changes.

Clinical photography is also helpful to monitor changes. A skin biopsy – small sample of skin tissue – may be taken for examination in a laboratory.

## **How is Parry-Romberg syndrome treated?**

The condition itself may be treated with strong anti-inflammatory medicines in the early phases. These include steroids and other medicines such as methotrexate.

As Parry-Romberg syndrome can affect various areas of the skull and face, treatment is best delivered at a specialist centre where a multidisciplinary team approach can be taken. The multidisciplinary team will usually comprise craniofacial (skull and face) surgeons, maxillofacial surgeons, ear, nose and throat (ENT) surgeons, ophthalmologists (eye specialists), plastic surgeons, dermatologists, dentists and

orthodontists, with other specialists brought in as needed.

Treatment to improve appearance is usually suggested when the condition is in its stable phase and growth has finished in adolescence.

Reconstructive surgery to improve the appearance of the affected side may involve transfer of muscle and bone from elsewhere in the body. Sunken areas can be improved using liposuction and fat transfer. Dental surgery may be needed to remove any malformed teeth. Some children and families benefit from psychological input at various stages throughout childhood and adolescence.

## **What is the outlook for children and young people with Parry-Romberg syndrome?**

The outlook for children with Parry-Romberg syndrome is good with no long term impact on bodily functions such as breathing, vision and hearing. The majority of children grow up to lead a normal life, working and raising a family.

## **Further information and support**

Headlines – the Craniofacial Support Group – is the main support organisation in the UK for families of children and young people affected by a craniofacial disorder. Visit their website at [www.headlines.org.uk](http://www.headlines.org.uk)

Changing Faces is another organisation that offers help and support to anyone living with a condition that affects their appearance. Visit their website at [www.changingfaces.org.uk](http://www.changingfaces.org.uk) or telephone their helpline on 0845 4500 275.