

Growth hormone treatment for children with PWS

Introduction

The decision to start your child on Growth Hormone (GH) can feel overwhelming, and parents often have a lot of questions around its use and how it is administered. There have now been many research trials showing clear benefits for children with PWS, and these include:

- Increased growth (height)
- Improved motor development, muscle strength and tone and better body composition (ie more muscle mass and less fat mass)
- Improved tolerance for exercise
- Improved bone health
- Improved respiratory function

GH is usually given in the form of a daily injection, and parents are offered full training in how to do this correctly.

Should every child with PWS have GH treatment?

It should be considered for every child with PWS, and the NHS will provide treatment if their endocrinologist recommends it, and has confirmed it is safe to go ahead. However, GH treatment is not compulsory and some families decide not to have it for their child.

There are some medical reasons why treatment with GH cannot be started:

- A significant illness in addition to the PWS
- Severe obesity
- Severe breathing problems
- Untreated central and/or obstructive sleep apnoea
- Uncontrolled diabetes

At what age should GH treatment start?

It is recommended by our PWS Specialist Clinics that GH is started by the age of 12 months.

How to obtain GH treatment for your child

GH treatment is always set in motion and monitored by a specialist – generally a paediatric endocrinologist. However, the actual steps involved in this process can widely vary between different regions and hospitals. GPs generally prescribe the GH on the instruction of the specialist, but do not usually initiate treatment.

The generic process is as follows:

- A specialist decides when to start GH for a child with PWS. Specific tests to prove GH deficiency are not required, but it is essential to prove the diagnosis of PWS by genetic analysis.
- The specialist who initiates the growth hormone will organise various tests prior to the start of GH, such as a sleep study and blood tests.

- Parents decide which GH preparation to use based on their own personal preferences and the devices available – the specialist should provide information and guide them.
- The specialist organises training to the parents and if appropriate to the patient in the home administration of GH. The training is usually provided by a specialist nurse or by the GH company itself.
- Depending on the local processes, the specialist or the patient's usual GP provides the prescriptions for GH on a regular basis. Most GH companies provide additional services like collection of the GH prescription from the hospital or surgery, and direct home delivery of the GH.

Why should your child have a sleep study?

A sleep study is used to determine if there are any underlying respiratory issues, and it's really important that every child has a study carried out before commencing GH treatment. Usually involving an overnight stay in hospital, it is painless and doesn't have any side effects for your child. It will involve placing ECG pads on your child's chest, a probe on their toe or finger and possibly a stretchy band with a sensor across their chest. On occasion, a sleep study may show that tonsils or adenoids need to be removed – if this is the case, a second sleep study will need to be carried out after surgery before commencing GH treatment.

Monitoring treatment

Your child should be monitored every 3-6 months to ensure that the treatment is both safe and effective. This can involve blood tests to look at levels of growth hormone, and repeat sleep studies.

Weight management during GH treatment

Growth hormone treatment does not change the problems of increased appetite in PWS, and is not a solution to weight gain. Children with PWS need less calories, approximately 60 - 80% compared to children without PWS, and regular exercise is really important. GH treatment will help develop muscle strength, making exercise not only easier, but more beneficial.

Length of time for treatment

Unless there are reasons why GH is no longer appropriate, treatment should continue until your child has stopped growing. Some adults with PWS are being offered GH treatment, although at the moment there is less research evidence for this and no NHS guidance for treatment.

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