The BSPED is one of the affiliated speciality groups of the Royal College of Paediatrics and Child Health.

The society aims to improve the care of children and young people with endocrine disorders or diabetes mellitus, by bringing together professionals from a range of disciplines.

This leaflet has been written by members of the BSPED & reviewed by the Clinical Committee. It is designed to giveyou some general information about your child's condition and treatment. Your child's doctor or specialist nurse will be able to answer any further questions you have about your child.

Date of first issue: 01/06/2018
Last review date: 22/05/2025
Review date: 23/05/2029

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British Society for Paediatric Endocrinology and Diabetes

Arginine Vasopressin Deficiency (Central Diabetes Insipidus)

Information for patients, parents and carers

What is a hormone?

Hormones are chemical messengers. They are made in glands and travel round the body in the bloodstream. Hormones affect how other organs in the body work.

What is the pituitary gland?

The pituitary gland is a pea-sized gland which lies deep within the brain and controls many body functions. It is made up of two parts (lobes). These are called the anterior (front) and the posterior (rear) lobes. The posterior pituitary makes a hormone, called antidiuretic hormone (ADH, also called vasopressin). Vasopressin controls the fluid balance in the body.

The amount of vasopressin secreted by the pituitary is determined by sensors in the brain. These sensors monitor the concentration of the blood and make adjustments for this:

- If you are becoming dehydrated, more vasopressin is made. This helps reduce the amount of fluid in your urine.
- If the blood is too dilute from drinking too much, less vasopressin is made and the kidneys pass out more urine.

What is Arginine Vasopressin Deficiency?

In arginine vasopressin deficiency (or cranial diabetes insipidus) not enough vasopressin is made. This leads to a lot of water loss in the urine. This causes dehydration, which makes the child feel thirsty and drink more fluids.

In a small number of children, the part of the brain that controls thirst is also damaged and so the child does not feel thirsty, even when they are very dehydrated. If this is not recognized and the child is not given enough fluids the child may quickly become very unwell.

Arginine vasopressin deficiency (previously known as diabetes insipidus) is different from diabetes mellitus. In diabetes mellitus, the problem is with high blood sugars, which causes excessive urine output and thus thirst. This is easily tested for by measuring sugar levels in the blood and urine.

Why does it happen?

Arginine vasopressin deficiency happens when the posterior (rear) pituitary gland stops making enough vasopressin. This can be for many reasons. Sometimes the child may have a medical condition that damages the posterior pituitary such as histiocytosis, sarcoidosis, meningitis, head injury or a tumour. It can also be associated with brain malformations that affect all the hormones in the pituitary gland.

Occasionally it may be due to genetic conditions in the family. Sometimes we cannot find a cause and then it is called idiopathic arginine vasopressin deficiency.

How does it affect the child?

A child with arginine vasopressin deficiency is very thirsty and passes a lot of urine. The symptoms depend on how much vasopressin the child can make. If they can still make some, the child may just get up at night to drink and pass urine. If they can make very little, they can be so thirsty they will drink from anywhere, which may include vases, taps and puddles.

When these symptoms are first noticed many parents and professionals naturally assume this is a behavioural problem and try to restrict the child's drinking. If a child has arginine vasopressin deficiency, this would be very upsetting for a child and should prompt families and professionals to consider further assessment.

Usually, affected children remain well if they can drink enough, but if they cannot drink sufficiently, for instance if unwell, they may become seriously poorly. If treatment with fluids is not given the child may die.

How is it confirmed?

Testing for arginine vasopressin deficiency involves measuring and comparing how concentrated the child’s blood and urine is.

If arginine vasopressin deficiency is not strongly suspected, your doctor may check blood and urine concentrations first thing in the morning. If the morning urine is concentrated, this can often exclude the condition, especially if there has been no fluid intake overnight. In this case the urine would usually be yellow in colour.

If arginine vasopressin deficiency is strongly suspected, the doctor may arrange a special test, called water deprivation test. During this the child is not allowed to eat or drink for up to 8 hours while samples of blood and urine are taken regularly to check how concentrated they are. This can be unpleasant and is potentially dangerous if not done properly so must only be performed in a specialist centre.

If arginine vasopressin deficiency is confirmed then the child will need to have more tests, including an MRI scan of the pituitary, to help find the cause.

How often does this happen?

Arginine vasopressin deficiency is rare, only developing in about 1 in 25,000 children. Less than 10% of arginine vasopressin deficiency passes from parents to children. Of the genetic causes, the overall frequency in the general population is estimated to be 3 cases per 100,000 population.

How is it treated?

The missing vasopressin can be easily replaced with the synthetic hormone DDAVP (Desmopressin). This may be as tablets, melts, nasal spray or injections. The effect is usually noticed immediately as the child is less thirsty and often stops getting up at night to pass urine and drink. DDAVP should be started at a low dose, which is adjusted over several days or weeks. This may be started in hospital and children usually need 1-3 doses each day. Your child will need regular blood tests to help the doctors know how much your child needs.

When the treatment is stable then your child will only need blood tests every few months, after any dose change or if they are unwell.

If the dose of DDAVP is not enough for your child or is missed then they will be very thirsty and pass more urine.

Having too much DDAVP can stop urine from being passed and increase the fluid in the body which is dangerous. If this happens then you should miss your child's next dose and contact your medical team for advice.

How long will this treatment be needed?

This depends on the cause but usually arginine vasopressin deficiency is a lifelong condition. Your clinic doctor or nurse will be able to give you more information about your child.

Are there any long-term problems?

With proper medical care and management, the symptoms of arginine vasopressin deficiency can be very well controlled.

Arginine Vasopressin Resistance

When the kidneys are unable to respond to vasopressin, this is called arginine vasopressin resistance. It has the same symptoms as arginine vasopressin deficiency but giving DDAVP (desmopressin) treatment does not work. There is usually a genetic cause and it is more common in boys than girls. This is a different condition and is managed by renal (kidney) doctors.

Suggested sites for further information

www.pituitary.org.uk

www.eurospe.org/

www.apeg.org.au