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## **Information Sheet**

# Precocious puberty associated with spina bifida and hydrocephalus

#### Normal puberty

Puberty is a complex developmental process which ends with sexual maturity. The time the process begins in any child depends upon many factors. Some of these are hormonal, some are genetic and others are environmental. The whole process works a little like a chain reaction and it is coordinated by two bodies within the brain, the hypothalamus and pituitary gland.

Puberty brings a number of different changes to the body. In boys, the change begins with sexual development and is followed by the development of pubic hair, acne, increased growth, increased strength, deepening voice and skeletal maturation.

In girls the changes begin with breast development and finish with menstruation. Along the way there is increased growth, skeletal maturation, body shape changes and uterine development.

Puberty usually extends for about two years for both boys and girls.

#### **Precocious puberty**

Precocious puberty is when the right things happen in the right sequence but at the wrong time. Sometimes things can happen in the wrong order or sometimes puberty may begin and then stop. These are not precocious puberty.

Nowadays the onset of puberty is not considered premature unless it occurs before the age of 8 in girls and 9 in boys. However, it does depend on a number of factors such as a person's race. For instance, Afro-Caribbean children tend to reach puberty earlier than Caucasian children.

Diet and nutrition also affect the timing of puberty. Girls today reach puberty 2-3 years earlier than they did 100 years ago. This is thought to be largely due to better nutrition today.

#### Spina bifida, hydrocephalus and precocious puberty

Children with spina bifida and hydrocephalus or hydrocephalus without spina bifida usually go through puberty earlier than other children. On average, boys with spina bifida and hydrocephalus reach puberty 1 year ahead of their peers and girls, 2 years ahead of theirs. Children with spina bifida but without hydrocephalus don't seem to reach puberty earlier.

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The reasons for this are not well understood, but it is thought that the alteration to the development of the brain associated with hydrocephalus affects the hypothalamus and pituitary gland and disrupts the timing of the onset of puberty. Other children with central nervous system disorders, brain tumours, meningitis and trauma are also more likely to have an early onset of puberty.

#### What problems are there

There are a number of problems likely to follow from or be associated with precocious puberty.

#### 1. Short stature

Before puberty a child usually grows at a steady rate, adding about 5-8 cm. per year to his or her height. Upon reaching puberty, the child undergoes a growth spurt. During this time, he or she grows about 10-15 cm. per year. While the sex hormones accelerate growth, they also speed up the process that ends growth. Children who start puberty prematurely are tall for their age, but since their skeleton matures and growth stops at an earlier age than normal, they don't reach their full height potential as adults. There are other factors involved with spina bifida which contribute to short stature, but adults with spina bifida, on average, are about 30 cm. shorter than the average person without spina bifida.

#### 2. Behavioural changes

Behaviour may change to that typical of an adolescent. Some girls who start puberty prematurely go through periods of moodiness and irritability, much like teenage girls. Some boys become more aggressive than their peers and develop a sex drive. On top of the selfconsciousness and lower self-image involved with being a child with a disability, being physically more mature than their peers is another issue many children are selfconsciousness about.

#### 3. Teasing

Teasing from other children about sexual development can also be a problem. Such teasing is especially common for girls who develop breasts. Parents can help by acknowledging that the teasing is upsetting and by helping children find a way to deal with it.

#### 4. Compounding social problems

Children with spina bifida often feel isolated and rejected socially. This is particularly true when, because of their learning disabilities, they have not been able to learn social skills appropriate to their age. Having another set of problems which leads to age-inappropriate behaviour adds to the problem.

#### Diagnosis

A number of medical investigations can be undertaken to assess what is happening in the child's body.

- Blood tests can be done to look for elevated levels of sex hormones and pituitary hormones.
- An X-ray of the hand can be performed to see how advanced the bone age is.
- In girls an ultrasound can detect enlargement of the uterus.
- The child's rate of growth can be measured.

#### **Treatment options**

- 1. The first option is to treat with a drug such as Medroxy-progesterone. This drug has been used to suppress activation of the pubertal process. However, it is not fully effective in inhibiting bone maturation or in improving adult height. It can also have side effects such as weight gain, moodiness and depression.
- 2. The second option is to treat with LHRH, another name for GnRH (For more information,

see the bottom of this sheet under *What Triggers Puberty*). Higher levels of this hormone in the body switch off the pituitary gland. No side effects for this treatment have been noted, but it is very expensive.

3. The third option is to do nothing. The side effects of this may be the problems noted above, however these may not affect all children to the same degree. The decision to treat or not to treat has to be arrived at with all the available information and must be made by the doctor and parents based on the diagnosis, prognosis and any other associated factors.

To be effective, treatment (if it is to be given) must begin early in puberty. For instance, it would be too late to treat a girl who has begun menstruation. Because menstruation occurs at the end of puberty, the changes that would be deferred by earlier treatment would already have taken place.

#### Summary

Children with spina bifida and hydrocephalus frequently have an early onset of puberty. Investigations carried out by a paediatric endocrinologist are aimed at confirming the diagnosis and giving an estimate of the prognosis in terms of the pace of future development and final height.

An added problem for children with spina bifida and precocious puberty is that the early rapid growth can increase the likelihood of tethering of the spinal cord. See the SBH Queensland Information Sheet on *Tethered Spinal Cords* for more information.

### What triggers puberty

The hypothalamus, a gland in the centre of the brain, releases a hormone (gonadotropinreleasing hormone GnRH) which in turn prompts the pituitary gland to secrete another set of hormones. These are called gonadotropins. The two main ones are luteinising hormone (LH) and follicle stimulating hormone (FSH).

In turn the gonadotropins induce the development of the ovaries and testes, which then release the gonadal steroids. These are the ones that most people have probably heard about.

Testosterone is secreted from the testes in males and oestrogen and progesterone are secreted from the ovaries in females.

The whole process is regulated a bit like a thermostat in the refrigerator. When the fridge gets too hot, it turns the motor on and when it gets too cold, it turns it off.

The brain senses the level of steroids in the blood and when they get too high, it decreases the production of GnRH which decreases the LH and FSH which then signals the gonads to slow the production of steroids.

When the level drops, the opposite happens. GnRH production is increased; LH and FSH increase and the gonads produce more steroids.

In children the hypothalamus is very sensitive to the level of gonadal steroids in the blood, and not much is produced. But when they reach puberty, the hypothalamus becomes less sensitive and so the levels rise. It is as if the thermostat in the fridge has been turned up.

The exact cause of this change is not completely understood, but it is known that genetic and environmental factors are involved.

In girls another important change occurs. The hypothalamus matures so that there is a positive feedback system instead of a negative feedback one. That is, an increased level of oestrogen in the blood leads to an increase in LH and FSH which leads to a greater level of oestrogen and so on. It is this surge in LH and FSH, which triggers the female's first ovulation. From then on, gonadotropins are released in monthly surges, which regulate the cyclical timing of ovulation and the menstrual cycle.

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