Puberty

in young people with disabilities

Puberty, contraception and hormonal management for young people with disabilities





Contents

Page	Section
1	Introduction
	Puberty – what is normal?
2	Normal puberty for girls
3	Normal puberty for boys
4	Early (precocious) puberty
	Early puberty in girls
	Early puberty in boys
7	Delayed puberty
8	Treatment of early or delayed puberty
	Early puberty in girls
	Early puberty in boys
	What treatment is available for early puberty?
9	When does delayed puberty need to be treated?
	Delayed puberty for girls: how is treatment given?
10	Late puberty for boys
	How is treatment given?
	Early and late puberty: key points
11	Menstruation and contraception
	Commonly asked questions for girls who have disabilities about menstruation and contraception
	Menstruation problems
	Contraception options
13	Other reasons for the use of contraception
14	Sexual function and fertility
	Sexual function
	Fertility
15	Notes
16	References

Written by A/Professor Margaret Zacharin D Med Sci FRACP, Endocrinologist, Endocrinology & Diabetes, The Royal Children's Hospital (RCH), Melbourne, with advice and guidance from Professor Dinah Reddihough, Developmental Medicine, RCH.

Introduction

Puberty, Contraception and Hormonal Management for Young People with Disabilities

This booklet will outline the normal process of puberty in males and females. Problems with the development of puberty that may be encountered for young people with disabilities will be discussed first, followed by available treatments

Puberty- what is normal?

The normal time of onset of puberty ranges from about 8-13 years in girls and 10-14 years in boys. Different ethnic groups and different communities vary widely in puberty onset. A pattern of both early or late puberty tends to recur in families.

Early pubic hair development is often seen in children born with structural brain problems such as cerebral palsy. This does not necessarily indicate onset of or progress through puberty. It is due to production of DHEAS, a hormone made by the adrenal glands located above each kidney. It does not need

treatment. However, early pubic hair can also be caused by more serious hormone problems, so it must be carefully investigated by a specialist.

Sometimes progress through puberty can be rapid and may require treatment. Frequently however, the early changes are minimal and progress may be extremely slow. Puberty can even switch off completely and the changes can disappear for a time.

Normal Puberty for Girls

The first change of puberty in girls is a small amount of breast development. This change is accompanied by a growth spurt in height. Pubic hair development can start around the same time.

What does it mean if my child has a period just after she starts developing?

At whatever age puberty commences in a girl, either early or later, the female hormone, oestrogen, has effects on all body organs. This includes the lining of the womb (uterus), which starts to increase in amount. If puberty then switches off spontaneously, the lining of the womb falls out, appearing as a small amount of vaginal bleeding. This may be interpreted by parents or carers as the onset of menstruation, however, it is not, This bleed simply indicates a loss of oestrogen. Further changes of puberty may not progress for months or years. This does not require treatment. This pattern may occur 2-3 times at the onset of puberty and then disappear completely as puberty progresses, re-appearing as normal menstruation about three years later.

Onset of menstruation (menarche) is usually 21/2-3 years after the commencement of puberty. Initially periods can be quite irregular, as with able-bodied girls. There may be only one or two periods in the first year. However, in some airls periods can be heavy. painful and very distressing and can occur every 3-4 weeks, lasting a variable amount of time, from 1-10 days.







Early changes of



Fully developed

Normal puberty for Boys

The first change of puberty is an increase in the size of the testes (testicles), followed by pubic hair and growth of the penis. In contrast to girls, a male growth spurt occurs towards the end of puberty, most commonly at 14.5-15.5 years. Early puberty is much less common in boys than girls. In situations where there is a disturbance of brain structure or function, this may trigger early onset of puberty.





Puberty



Fully developed



Early (precocious) puberty

Some children who have abnormalities of brain structure of any cause, may have early or precocious puberty. This type of problem is more common in girls but also occurs in boys.

Early puberty in girls

When puberty is early in a girl who has an intellectual and physical disability, parents have major concerns for the future. These range from fears of the child failing to understand her own changing body, concerns about onset of menstruation, differences from peers, concerns regarding rapid growth and height outcome, together with grief for a loss of childhood.

Parents have questions about contraceptive needs, menstrual management and possible unwanted exposure to sexual behaviours, either on the part of the girl or by others towards the child. Sometimes they may not feel comfortable to discuss these concerns openly with their doctor

All of these are very real concerns that need to be individually addressed. It must be emphasised that most girls who have early changes of puberty coupled with disability, do not suffer most of these problems. When necessary, effective treatment to switch off puberty is available.

Occasionally puberty progresses very rapidly, even from a very early age such as 2-4 years. Quite severe mood change can occur as the affected girl experiences hormonal fluctuations. If the child is nonverbal or has limited understanding, this can be particularly difficult for families and may only be detectable as increased agitation or crying, with no apparent source of discomfort being located.

Rapid breast development can be uncomfortable in any girl, particularly in the early phase of growth. Where a child wears a restraining seat belt or harness, there might be significant but unrecognised pain and tenderness where the growing breast or nipple rubs against the harness. Due care needs to be given to protection in this area.

Menstruation at an unusually early age can cause anxiety for parents and is occasionally difficult to manage for carers. It can also be associated with pain and agitation or increased crying in a nonverbal child. If this happens, treatment may be required to switch off periods.

Girls who have intellectual disability and early puberty may be more prone to masturbate in a public situation, due to a relative lack of conventional social inhibition. This can cause distress to families and may occasionally be a reason for intervention.

Early puberty in boys

Rapidly progressive early puberty is much less common in boys with disabilities than in girls, although onset that is slightly earlier than that for other children in a family is more common. Although it can occur in association with structural brain disorders, it may rarely indicate another new underlying disorder such as a cyst or tumour in the hypothalamic area of the brain. Thorough assessment is required for all boys with very early puberty.





Delayed Puberty

Puberty may be late or may progress very slowly in young people with severe disabilities. This may be due to the child being very thin or may simply be part of a general delay in development.

Parents of children with disabilities are often relieved if puberty is late. They may have concerns about the psychological and social aspects of puberty, particularly when a child has an intellectual disability and cannot fully understand the process of growing up. In particular, there are sometimes concerns about public masturbation or unusual sexual behaviours. These behaviours are uncommon. Rarely parents may even want to prevent the child from going through puberty at all. This is not a good idea, for several reasons.

There are a number of advantages to encouraging normal progress through puberty. Generally, the addition of sex hormone improves both physical and psychological maturity, even when a young person has a severe intellectual disability. Often parents comment that even minimal understanding and co-operation can markedly improve as the child grows up. Physical strength improves so that the voung person may be able to assist with their own lifting and position shifts. Mood is frequently improved and for most young people the overall experience is satisfactory.

If puberty is extremely delayed, sometimes a short course of sex hormone may be required, to stimulate normal ongoing development. This treatment is quite safe, with no side effects. Rarely, if puberty does not happen at all, it may be necessary to give hormone support until adult body size and appearance is reached. This type of intervention requires specialist advice and management.

Bones become thicker and stronger during puberty. Failure to do anything about helping, if puberty is severely delayed, can prolong the time of increased fracture risk that accompanies immobilization and a small bone structure.

Other Hormone deficiencies causing growth and puberty problems

Occasionally, children are born with midline abnormalities when the brain is being formed, or sometimes a traumatic brain injury may have resulted in damage to the hypothalamus or pituitary gland. In both situations, multiple hormone deficiencies may occur. Lack of growth hormone and thyroid hormone as well as the sex hormones of puberty all need medical tratment, to improve general growth and health. Rarely, another hormone (Prolactin) may be increased. If this happens a high prolactin may prevent puberty from progressing and may also need treatment.

Treatment of precocious or delayed Puberty

Treatment of early puberty in Girls

In the general population, when puberty is unusually early, families may seek treatment to reduce or reverse the effects of puberty and also to try to help preserve the greatest growth potential, to maximize adult height.

For children with disabilities who have early puberty, the situation may be different. The usual reason for treatment to be considered for girls with disabilities relates to heavy, frequent periods at any age, associated pain or serious parental distress, rather than height preservation.

It may not be an advantage to be taller if the young person will always require the use of a wheelchair.

For some families, being taller is important so this would also be considered, as for any child with early puberty. Although it is quite simple to switch off early puberty, treatment does not usually make much difference to final adult height.

Treatment of early puberty in boys

Precocious puberty in boys with disabilities is far less common than for girls but it can be treated in the same way, using exactly the same treatment methods as described for girls.

What treatment is available for early puberty?

There are two options for treatment, where needed.

GnRH agonist

This is a synthetic form of a small brain protein that acts to "flood" the receptors within cells and thus switch off puberty. It

is very safe and effective and helps to some extent, to improve final height outcome. The main disadvantage however is that it must be given by injection, usually every 1-3 months. It can be uncomfortable and frightening for a child, particularly if understanding is limited. It is also extremely expensive in some countries.

Progestogen

When achieving the greatest possible final height is not a concern, precocious puberty can be treated with a progestogen, a derivative of natural progesterone. It alters the lining of the womb and stops periods. Its action is less strong than GnRH agonist. Therefore, it is less efficient at maximizing the best final adult height outcome. This medication can be taken as a daily tablet.



When does late Puberty need to be treated?

The commonest cause of very late puberty is a very thin child with poor nutrition. Sometimes, even after late puberty has been treated, this type of young person often cannot maintain adult levels of sex hormone and long term hormone support is needed to maintain health and to keep bones as strong as possible.

Occasionally a structural brain abnormality may be accompanied by absence of the hormones that control onset of puberty. In this case long term treatment with hormone replacement treatment for either boys or girls will be necessary. It is rare.

Delayed or Late puberty for Girls

How is treatment given?

If puberty is very late (over age 14 years) it can be helpful to stimulate its onset with a short course of treatment. If the young girl is extremely thin or chronically unwell, this may be insufficient. It is sometimes necessary to take a child completely through puberty with hormone replacement treatment and then to take her off treatment, to re-assess whether her own body is able to continue to produce sex hormones unassisted.

To treat puberty in girls requires the use of oestrogen, the major female hormone. It can be given either as a tablet or as an adhesive skin patch, changed once or twice per week. The dose is slowly increased over 2-3 years, with addition of the other female hormone, progestogen, towards the end of the course, to allow a period intermittently. If, in the meantime. the development of the girl physically exceeds the changes expected for a given dose of medication, it can usually be assumed that spontaneous puberty is occurring and treatment can be withdrawn. The girl will need ongoing assessment to make sure that development continues to progress normally.

Once periods have been established, this can pose problems for a family and for the girl. Strategies for menstrual control may be needed (see below).

Late puberty for Boys

Puberty that has not commenced by age 14.5 to 15 years is usually considered to be late enough to treat. For a normal boy who has very late puberty, two to three intramuscular injections of testosterone, around 3 weeks apart, may be sufficient to prime the brain to commence puberty spontaneously. For a boy with a major disability and delayed puberty, as with girls, treatment may need to be longer, continued until the boy is fully developed. Then treatment can be stopped to see if the boy is able to sustain normal adult hormone levels without help.

How is treatment given?

Testosterone, the normal male hormone may be given as a capsule at first. This type of male hormone is very weak, but is strong enough to slowly introduce a boy to the normal hormone changes of puberty. If prolonged treatment is needed, other forms of testosterone can be used.

A gel is very effective but it needs to be rubbed into the skin 1-2 times every day. If a woman is administering testosterone gel. she must wear gloves because the hormone is rapidly absorbed through skin. A hormone containing skin patch can be used but it has a moderate risk of allergic reaction.

All puberty treatments are administered in slowly increasing doses, to mimic the normal stages of development, over about 3 years, if necessary, If permanent treatment is needed, a long acting intramuscular injection of testosterone is also available.

Other problems for boys with physical disabilities include possible problems with the descent of the testes. In boys who have spasticity associated with cerebral

palsy, testes are often found either high in the groin area or they may appear to be undescended. It is usually possible to coax the testes into the scrotum (sack) with a medical examination. However, the testes may continue to ride very high throughout adolescence and puberty. If this is the case, they may occasionally be compressed or squashed in the groin area. particularly if the boy has to sit in a chair all day. This can be a cause for a non verbal boy to feel very uncomfortable and to seem unsettled for no apparent reason. Consultation with a surgeon might be required under these circumstances.

If the testes are truly not properly descended, surgery is required, both to place them in the scrotum to reduce discomfort and to be able to see and feel the testes throughout life. A testis that was previously undescended may be more prone to abnormalities (including a risk of cancer).

Boys requiring long term treatment may continue with testosterone, as outlined above. Side effects are rare.

Early and late puberty: **Key points**

- If puberty is slightly early or late, treatment is usually not necessary
- If puberty is very early or rapidly pogresses, treatment is available for boys and girls, using GnRH agonist or progestogen See page 8)
- If puberty is very late or very slow, treatment is available for boys and girls. using small doses of male or female hormones

Menstruation and Contraception

Commonly asked questions about Menstruation and Contraception for girls who have disabilities

- What can be done to control periods?
- Does she need to have periods at all?
- What type of contraception is available for girls who have disabilities
- Who can use contraception
- When should it be used?

Menstruation problems

Girls who have either physical or intellectual disabilities or both, may have problems with management of regular menstruation. Difficulties of understanding, hygiene management and simply the physical burden of care required may cause parents, carers or the adolescent herself, to seek advice. A number of practical solutions are possible.

Continuous oral contraceptive pill is probably the most commonly used method to stop menstruation. However, it alters clotting factors. In an adolescent or young adult who has a chronic physical disability, the risk for vein thrombosis and / or lung clot may be higher than in a healthy girl. This risk may be increased with the use of the contraceptive pill, simply because the person sits for prolonged periods with flexed legs.

The "pill" may be used in a continuous fashion, by cutting off and not using the lactose (sugar) pills. In this way, periods should not occur at all. However, for some girls, "break through" bleeding can happen at unexpected times. Families may find this a problem.

For girls who have epilepsy and who require treatment with anticonvulsants, oestrogen is removed from the body more rapidly through the liver. A higher dose pill may be needed to control menstrual flow. This, in turn, may further increase the risk of clotting.

The "pill" is contraceptive as well as controlling periods. No pill is automatically right for every girl. Mood change or weight gain might occur with any girl who takes the "pill." Several types of "pill" might need to be tried before one is found that is satisfactory without side effects.



Various oral contraceptive pills

A progesterone bearing intra-uterine device

has proved very successful for use with young people who have physical or intellectual disabilities. It is a small plastic device that contains a type of progestogen that slowly leaches into the lining of the womb, drying and thinning the lining so that no periods occur. It lasts about 4-5 years. together with complete contraception during that time. It is changed by a gynaecologist every 4 years. The device requires a light general anaesthetic for insertion, in the adolescent. Insertion is limited by the ability to pass through a tight cervix. There is a possibility that it might fall out spontaneously, a risk that is common to all intrauterine

devices (IUDs). This type of problem could be detected by unexpected return of menstruation or by the finding of the device in a pad or nappy, if worn. This complication is rare.

Depo Provera is a long acting progestogen, given by intramuscular injection. It is very effective in stopping periods and is also contraceptive. However, it is extremely powerful and switches off the hypothalamic-pituitary-ovarian axis (message s from the brain to the ovaries). and completely switches off oestrogen in many patients.



An intrauterine device



Depo Provera

Lack of or loss of oestrogen reduces bone quality and increases bone fragility and fracture risk in all women. This is an extremely important problem. It is often unrecognised by parents and doctors alike. If this type of treatment is to be used, it MUST be given in combination with oestrogen, usually as a skin patch.

Implanon is a low dose, implantable progestogen that provides 3 years of contraception. Using local anaesthetic, it can be placed under the skin of the upper arm. Only a small portion of users lose their periods completely. Most girls have a very light but regular period and a few suffer frequent or continuous bleeding, necessitating removal of the device. Its use should be considered carefully in a girl who has an intellectual disability. Some girls may not tolerate the device that can be felt and may try to "pick" at it. Insertion of Implanon requires a large dose of local anaesthetic which is painful and frightening for girls who have limited understanding. It often causes weight gain and moodiness and is not usually recommended for gils with a disability.

Sexual function and fertility for young people with disabilities

Contraception for Girls

When an intellectual disability is present, parents and carers often have great fears for a teenage girl, in terms of even remotely possible unwanted exposure to sexual advance, usually in a situation when the girl is out of direct parental care.

The issue also arises if a girl who has a

milder disability may be uninhibited. In this case she may invite sexual advances or initiate sexual behaviour herself, without understanding the consequences.

Use of the contraceptive pill, Mirena IUD or Depo Provera plus oestrogen, is appropriate and allows protection for all girls. Choice may be determined by the type of care situation. For example, reliability of administration of a daily contraceptive pill when a girl is in respite care may not be accurate.

For girls with physical disabilities that are associated with extreme small stature. choice of contraception can be difficult. The use of the oral contraceptive pill in a dose suitable for an average sized female, may be excessive and might increase the risk of thrombosis. The lowest available dose of oestrogen should be chosen, but this may be associated with break through bleedina.

Even if a girl is extremely small, her womb (uterus) is usually a normal adult size. It may be possible to use a Mirena IUD. These girls require specialist gynaecological consultation and care.

Contraception for boys

Boys who have solely physical disabilities that are associated with extreme proportionate short stature may occasionally have a potential problem. Generally, small body size is usually not associated with small genital size but this can occur for some extremely physically small men. They should be aware that a variety of condom sizes are available if needed. These can be purchased from speciality shops.

Sexual function and fertility

Sexual function

For those young people who have either a purely physical disability or a mild to moderate intellectual disability, sexual function should be as normal, as for other members of the population, unless there is interference with nerve function. If a spinal abnormality or degenerative condition of nerves has caused a progressive or severe loss of nerve function, there may be a permanent loss of erectile function in boys. Detailed management of this problem is beyond the scope of this booklet. Specialist advice and management is available.

For girls with lower body paralysis or loss of function, sexual function is still possible, but an increased risk of urinary tract infection requires awareness and should be checked for regularly.

Fertility

After puberty, fertility is likely to be normal for most adolescents and young adults with a physical or intellectual disability. For girls with intellectual disabilities, some parents wish to make long term decisions on behalf of their daughters, to limit reproductive potential. With modern effective contraception being available to suit most circumstances, requests are not often made for hysterectomy, as occurred in the past. Under exceptional circumstances, if all other strategies fail, this possibility might need to be discussed with a specialist gynaecologist. In many countries hysterectomy can only be performed for affected girls after a decision has been made in a court of law.

For girls of extremely small physical stature, the possibility of pregnancy would need to be discussed in detail, before considering conception. Consultation with a respiratory physician as well as with an obstetrician is necessary. One of the major limiting factors for pregnancy under these circumstances is lung capacity in later pregnancy.

Sometimes, for a girl who has an extremely small body size, this may mean that a full term pregnancy is not possible, although achieving and maintaining a pregnancy up to 28-30 weeks gestation may well be an achievable target.

Notes

References

- 1. Zacharin MR, Puberty, Contraception and hormonal management for young people with disabilities Clin Pediatr (Phila). 2009 Mar;48(2):149-55.
- 2. Albanese A, Hopper NW Suppression of menstruation in adolescents with severe learning disabilities Arch. Dis Child 2007:92:629-632
- 3. Grover S, Menstrual and contraceptive management in women with an intellectual disability Ann N Y Acad Sci. 2008:1135:230-6
- 4. Quint EH, Menstrual issues in adolescents with physical and developmental disabilities MJA 2002; 176 (3): 108-110
- 5. Merck booklet, Problems of puberty. Part of 'Hormones and Me' series ed. Margaret Zacharin





