CF Guidelines - Growth & Puberty

Growth:

Infants with CF often grow poorly in the first year of life (especially before diagnosis), as nutrition is the principle factor in determining growth at this age. However catch-up growth usually occurs once treatment is established so that expected height is reached by 5 years. Growth through middle childhood is usually normal but a period of slow growth is often noted in the pre-pubertal period leading to a decline in centile rank round this age. It is often noticeable to the child and family as they do not have the rapid acceleration in growth expected at the onset of puberty, so seem to fall behind friends of a similar age. The majority of patients with CF show some retardation of growth in late childhood and this problem is exacerbated by the delay in onset of puberty that is commonly seen in CF. However the children tend to keep growing for longer so often regain their original centile. The final height of the majority of patients falls within the normal range although is usually less than expected from mid-parental height; only a minority is below the 3rd centile. The disease itself does not affect growth hormone secretion but growth problems are usually related to poor nutrition, recurrent infections and disturbed lung function. Optimising CF therapy is the correct approach to a falling height centile, there is no evidence that growth hormone injections are of benefit.

Patient monitoring and management:
Height (measured with a stadiometer) and weight should be recorded at every clinic visit (minimum ever 3 months) and plotted on the standard growth centile charts (do not circle the dots). In children under 2 years, head circumference (OFC) should be plotted if they presented with failure to thrive or if growth is still a concern. In adolescents, full pubertal assessment (Tanner staging) should be carried out at annual assessment. In those with delayed puberty a wrist radiograph should be done to assess bone age. If a patient is not managing to track an appropriate weight centile for height or is falling through centiles then action must be taken, including a full dietetic assessment. Often input from the clinical psychologist is vital, as feeding behaviour problems are common in young children. Supplemental feeds should be started early (see section 7). Referral to Dr N Bridges may be appropriate. Even if only to reassure the family that everything possible has been considered.

Puberty:

Puberty is often delayed in CF especially in those with nutritional problems. Delayed puberty is detrimental not only from a psychological perspective but also because of the negative impact that it has on bone mineral density. It is defined as no signs of puberty in a girl of 13 or a boy of 14.

First signs of puberty:

- Girls B2, (98% girls by 13.8 years).
- Boys 4ml testicular volume; 98% boys by 13.3yrs.

Pubertal Growth Spurt:

- Girls B2 – B3.
- Boys 10-12ml testicular volume

Assessment of Pubertal Delay:

- Measure height and weight.
- Tanner staging.
- Serum LH and FSH will exclude gonadal failure (when levels will be very

high) but are not helpful in predicting the time of onset of puberty.

Treatment of pubertal/growth delay:

There is no height benefit derived from delayed puberty. It is important to maximise nutritional status to stimulate pubertal development. However illness or poor nutrition may cause a pubertal delay which persists even after their resolution. Referral to Dr N Bridges is mandatory in those requiring active treatment.

Treatment aims:

- Psychological and social.
- Bone density – CF patients are at risk of reduced bone density and osteoporosis. None density increases during puberty as a result of sex steroid action, so it is important to treat pubertal delay and maintain normal sex steroid levels in adult life.

Reasons to delay treatment:

- Pubertal growth is reduced by illness, poor nutrition and corticosteroid treatment and it is important to try to correct these. However, if the clinical situation is not likely to change, then treatment of pubertal delay may be considered, accepting that optimal pubertal growth may not be possible.

Treatments available:

- Sex steroids. Dose is gradually increased over 2-3 years mimicking puberty.
- Girls - Start oral dose of 2 mcg ethinyloestradiol once daily. At doses of 15 mcg ethinyloestradiol commence cyclical progesterone.
- Boys - Start 50mg testosterone esters (Sustanon) IM, every 6 weeks. Caution is required as it can cause hepatotoxicity, so use carefully in established CF liver disease.

Oxandrolone is a weak androgen with growth promoting effects. For use in boys in early puberty who are some way from their pubertal growth spurt. It can stimulate an increase in growth rate without impairing final height. Dose is 2.5mg oral, od for 3 months. Oxandrolone can be also used to stimulate a short term increase in prepubertal growth in males and females. Dose used is 1.25mg od for a maximum of 6 months, longer courses may advance bone age. The growth stimulating effect of oxandrolone is inferior to sex steroids and it should not be used as an alternative.

Stopping treatment:

- Stop treatment if endogenous function starts. Otherwise, stop treatment for a short time at the end of growth to reassess endogenous function. If treatment is continued, it is important to check that adult sex steroid con centrations are reached.

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