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**CAH**  
**MANAGEMENT THROUGH**  
**CHILDHOOD/ADOLESCENCE**

*Transcript prepared by Sue Elford & checked for accuracy by Dr Stanhope*

## Management Through Childhood/Adolescence

Dr Stanhope spoke about the problems in adolescence and how important it was to get the management of glucocorticoid and mineralocorticoid treatment right in childhood in order to minimise those potential problems. If too much hydrocortisone is prescribed, growth will be slow and it can lead to obesity, hypertension and osteoporosis.

He said that he generally doesn't perform biochemistry tests if he can see a child is progressing well, although he does regularly check blood pressure and organises a bone-age x-ray (every 12-18 months). The latter confirms that bone-age is in line with chronological age and growth is appropriate. A blood test may be taken (approximately once a year) to measure renin (in salt losers), which will show whether the fludrocortisone dose is appropriate. If well managed throughout childhood there is no reason why this should change in puberty/adolescence. It is vital to obtain the correct height to weight ratio at early adolescence.

Puberty often comes a little earlier in CAH (especially with the non-salt-losers who are generally diagnosed between the ages of 3-7 years and who already show signs of precocious puberty). It can occur early in salt-losers too though, particularly if the bone-age has advanced. When puberty occurs too early, there is a treatment, which is effective and involves injections, which will suppress puberty. These are usually given on a monthly or three monthly basis until the child reaches a more appropriate age, when puberty can be entered normally.

The main problem seen with adolescents is non-compliance. If a teenager misses tablets on a regular basis, they lose control and over a period of time this has its effects. Losing control can affect weight and if an adolescent becomes overweight it can be difficult to get them back on track. The problems in adolescents may be psychological. Non-compliance is not just a problem with CAH but applies to other long-term medical conditions too. It is relatively common behaviour for a teenager to rebel and not to want to be reliant on medication. It is therefore important that they are aware of the risks involved! The problems can also be medical. Growth is dramatic during puberty and the doctor looking after the child has to be one step ahead as if an adolescent is starting to grow out of their dose, they need to increase it at the first signs and not leave it until the next appointment six months later, as control could be lost by then.

In children with CAH theoretically a normal replacement cortisone dose is required but in order to suppress the excess androgen production, you have to give extra hydrocortisone. However, if you give too much, the unfortunate side effect is obesity. If this is a problem, a blood test will show if a child is over suppressed. Some centres do saliva tests, which are good for screening but blood tests are more definitive. Some children experience headaches if the dosage is not quite right. This can be due to cortisol deficiency, although the total daily dose may not be insufficient. Adjusting the timing or the amount of the dosages may help.

Hydrocortisone lasts in the body approximately four to six hours. Ideally it should be given four times a day. This isn't very practical though and could make compliance difficult for anyone. Therefore thrice daily administration is usually recommended and Dr Stanhope suggests the first dose should be given early morning on waking, preferably before getting out of bed. The morning dose he recommends represents 50% of the total daily dose. There should be a second dose taken around lunchtime and a third in the evening, before bed. In many centres in the USA, they recommend 2am as the optimum dosage time (as this is when there tends to be a surge of androgens). That isn't considered necessary by most consultants in the UK. Patients differ in how they metabolise hydrocortisone and in puberty they can clear it at a faster rate. As long as patients are monitored adequately control is still manageable.