

Growth in Noonan Syndrome (2004)

Dr. M.O. Savage of St. Bartholomew's Hospital, along with other groups, has been involved in a joint project, of the use of growth hormone in NS and presented his data on this trial study.

In NS 50% of children have short stature below the 3rd centile, clearly showing that this is a problem. The question had been addressed as to whether or not these children could be helped. Data collected on 6 children four years ago had shown that there was a strong possibility of qualitative abnormality rather than quantitative abnormality of growth hormone secretion in NS. They showed a normal growth hormone response to a provocative growth hormone stimulation test and a definite increase in their growth rate over a period of 1 year which reinforced the idea that this should be looked at on a formal basis.

In conjunction with Serono Laboratories a formal clinical trial was set up nationally, for maximum recruitment as cases were so scattered. The dose used is that shown to be effective in Turner Syndrome.

Aim: To study the efficacy of growth hormone treatment in children with NS over a period of 1 year. To document changes in cardiac morphology during the study period.

Criteria: Children should be between 5-14 years old.
Should have short stature but not necessarily growing slowly.
Every child should have been seen either at St George's or by one of Professor Patton's team to confirm NS diagnosis.
Should have normal maximal left ventricular wall thickening on 2D Echo.

Exclusions: Children with severe hypertrophic cardiomyopathy.
Good general health.
Standard growth hormone stimulation test to ascertain that they were not growth hormone deficient.

Treatment: Daily subcutaneous injections of growth hormone.

Measured: Height, ECG, Blood tests – at 3 months.
Bone age – at 6 months.
Echocardiogram – at start and end of period.

Recruited: Of 30 patients recruited there is data on 22, 7 female and 15 male in the age range 9.7 - 0.6 with a bone age delay of +/-2 years, height of below 3. 3rd centile, growing fairly slowly with normal growth hormone to a provocative test.

Protein: With cholesterol as a precursor a growth rate may be related with lipid synthesis. The rate of synthesis of cholesterol is regulated by the enzyme HMG-CoA reductase. The rate of synthesis of cholesterol is regulated by the enzyme HMG-CoA reductase. The rate of synthesis of cholesterol is regulated by the enzyme HMG-CoA reductase.

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