FACTSHEET #6
PUBERTY & GROWTH

Noonan Syndrome Association

The age at onset of puberty is often delayed and short stature is common in Noonan syndrome, although adult height is not always affected. In boys there may be problems with the testicles descending (cryptorchidism) and reduced fertility in adult life.

Puberty

The average age of the onset of puberty is delayed in people with Noonan syndrome compared with the general population:

- 35% of boys enter puberty after the age of 13·5 years
- 44% of girls enter puberty after the age of 13 years.

Fertility is not impaired in women with Noonan syndrome.

In a significant number of boys with Noonan syndrome one or both testes may fail to descend into the scrotum (cryptorchidism). This should be corrected by early surgery which helps to reduce the incidence of fertility problems with lower sperm counts later.

Growth

At birth, the baby's weight and body length are usually normal. But as the growth spurt that comes with puberty is often delayed or longer, short stature is common during the age of normal puberty. The adult height of people with Noonan syndrome is not always affected – but on average it is reduced.

It is useful to monitor growth and refer to the growth charts for Noonan Syndrome where possible. If growth is significantly delayed it may be helpful to have a referral to a specialist paediatric endocrinologist.

Although the level of growth hormone is usually normal, children with short stature in Noonan Syndrome will usually respond to treatment with regular growth hormone injections. There is some controversy about this as the treatment will be required over several years and is expensive. Most children with Noonan Syndrome will get an increase in height from the treatment which may be very helpful in boosting the child's self-confidence. However there is still some debate about how effective the growth hormone treatment is in increasing the final height.

At the present time growth hormone is only available through a specialist paediatric endocrinologist. The NHS has not recommended growth hormone for all children with Noonan Syndrome but the position on this is currently under review. The presence of an underlying cardiomyopathy may be a contraindication for growth hormone as there have been reports of cardiomyopathy increasing on growth hormone treatment

Other growth-related issues include spinal deformity, chest deformities ('pigeon chest' or a sunken chest), widely spaced nipples, forearms that angle away from the body to a greater degree than normal when fully extended (cubitus valgus), and knock knees.

REFERENCES/FURTHER READING

Shaw AC, Kalidas K, Crosby AH, Jeffery S, Patton MA. The natural history of Noonan syndrome: a long-term follow-up study. Arch Dis Child. 2007 Feb;92(2):128-32. doi: 10.1136/adc.2006.104547.

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