

## **INFORMATION FOR TEACHERS**

### **What is Noonan Syndrome?**

Noonan Syndrome was first named by Dr Jacqueline Noonan, an American paediatric cardiologist, who in 1963 recognised that an unusual congenital heart defect, was often accompanied by short stature and a characteristic facial appearance. The Syndrome itself is a complex one with many different features recognised. Most children with Noonan Syndrome do not possess every feature, but each characteristic can be expressed to a differing degree in an individual child. Thus, most children are mildly affected and cope well in mainstream. Others are more severely affected in one area or in several, and require provision at an MLD school. Each individual has differing needs. Many children are at Stage 3 of the Code of Practice. Some, at Stage 5, have support from an NNEB or Special Support Assistant (SSA). It is not known how many children are receiving support nationally.

The incidence of Noonan Syndrome is between one in 1,000 to one in 5,000 of the population. Half of all cases run in families. Recently genetic testing has been developed as a gene for Noonan syndrome was discovered (PTPN11). There are more genes to identify and so there will be some children in whom the diagnosis is still made clinically.

### **Physical Effects**

- Abnormal birth weight
- Early sucking and feeding problems
- Dental problems
- Poor sleep patterns
- Major delay in the developmental milestones of sitting, crawling, walking, talking, etc
- Low immunity to colds, coughs, chest infections, tonsillitis, etc
- Susceptibility to easy bruising and profuse bleeding

*Facially, children have a recognisable appearance-*

- Low set ears
- Drooping eyelids
- Large downward slanting eyes
- Eyes spaced wide apart

- Broad, flat bridge of the nose
- Short neck
- Excess skin on the back of the neck
- Low hairline

*Other major physical features include-*

- Short stature
- Underweight
- A "pigeon-chest"
- Undescended testes in boys
- Cardiac abnormalities – the most common being the narrowing of the pulmonary valve which takes blood from the heart to the lungs

Noonan Syndrome may be the second most common Syndrome after Down's Syndrome in the incidence of congenital heart conditions.

Obviously it is wise for the teacher to check with the parents what the implications of such a condition are for a child terms of tiredness, PE etc.

Many of these problems will require frequent visits to clinic or hospital for treatment or surgery. These unavoidable absences disrupts the child emotionally, and academically.

### **Sensory Problems**

- *Vision*

Some children have a squint and attend an Orthoptic Clinic. Short-sightedness may also be present. These problems can be corrected by glasses.

- *Hearing*

This may fluctuate due to glue ear (the build up of fluid in the ear). Grommets may need to be inserted. Occasionally there may be more severe nerve deafness.

### **Speech and Language Delays**

Early hearing loss plays a role in delaying the onset of speech. Many children with Noonan Syndrome require speech therapy as early as possible to deal with delays in articulation, or in semantics such as using words inappropriately. Delayed speech also affects the acquisition of literacy, so children often find it a real struggle to learn to read.

## **Co-ordination Problems**

Input from a physiotherapist or occupational therapist is often required to treat these-

- Poor muscle tone
- Instability at the shoulder or pelvic girdle
- Mild spinal curvature
- Hyperextension of some joints

Poor co-ordination and delays in gross motor development result in early difficulties such as delays in toileting skills, or dressing themselves and their dolls, as well as difficulties in fastening button, zips, hanging up coats or learning to tie up shoe laces. In school, clumsiness is apparent.

The drawing skills of children with poor body control and awareness may reflect their difficulties. Their art is immature, with evidence of poor joins and inadequate human figures. They will avoid drawing and colouring so exacerbating the problem through lack of practice.

At the level of fine motor control, a wide carrying angle of the elbow, small hands, and a low set thumb, all produce major difficulties for pincer grip and pencil control. Thus handwriting, drawing, tracing and copying are extremely difficult for children. These problems can be alleviated to a certain extent by providing the child with pencil grips or 'Handhuggers' from Berol. Lined paper is essential and a sloping desk (such as the medieval scribes used) will support the writing arm.

Advice given in the various books written by Jean Alston are of great help here. Older children may well need a laptop. Young children will find certain types of manipulative play like threading beads, using peg boards, moulding plasticine, baking and linking manipulative toys like Lego extremely difficult. SENCOs would need to refer to an Occupational Therapist for specialised equipment, if necessary.

## **Cognition and Intelligence**

There are now a few studies available on cognition and intelligence in Noonan Syndrome. There is quite a wide range of intelligence with some children going to University but there is an over-representation from

children with mild intellectual deficit. In Noonan Syndrome a common pattern of cognitive deficits has been observed.

These are-

- Short concentration span
- Distractibility
- Difficulties in sequencing
- Poor short-term memory retention
- Inability to select relevant information from trivia
- Poor visual/spatial skill
- Poor reasoning ability

All these defects will affect every aspect of the curriculum. Repetition and reinforcement will be required in many skill areas. In particular, routines and procedures, e.g. in maths, will need much over-learning. Children will find it difficult in learning multiplication tables, and in the instant recall of number bonds.

Interestingly, many of the above features are also found in Dyspraxia, Specific Learning Difficulty and ADHD. Experienced teachers will be able to adapt some of the techniques already in their repertoire for use with children who have Noonan Syndrome.

### **Personality and Behaviour**

Research has suggested some of the behaviours are challenging and frustrating for parents and teachers to deal with. Children with Noonan Syndrome are socially immature, preferring to play with younger children, but they can be protective and caring towards them. They are not accepted by their peers, although a contributing factor here could be their poor physical skills. Their immaturity and lack of common-sense means that they need much more adult supervision than would normally be given to a same-aged child. They take much longer to acquire road sense.

Children with Noonan Syndrome can be stubborn and obstinate with an action or idea long after it's run its course.

Behaviour may be repetitive – beware of the library shelf tidier! They dislike change in their daily routines, so the arrival of a supply teacher can be upsetting for them. Tantrums may appear without an obvious cause.

They are egocentric and heavily demanding of attention. This may make them appear over-friendly or even over-familiar. But they also demand attention from peers too, and this may be another reason for their social isolation – they don't seem to perceive in sharing their possessions with others and can be obsessive about the care of these.

On the plus side, the determination and perseverance they can show can be a great asset.

### **School Policy Classroom Management**

We live in a society that clues height. So the twin problems of being short and having learning difficulties may result in a child with low self-esteem and consequently poor school performance. Class teachers should ensure that the classroom is physically accessible to a short child so that he/she does not have to constantly ask for help in reaching things or putting them away. Physical prowess has different values according to the age of the class. It becomes increasingly important with age. Failure and ridicule are likely to follow the child's attempts at PE and games. He/she is always the one who comes last in races and is last to be picked for teams. Sports days are a nightmare for the physically-challenged! Children with Noonan Syndrome are also targets for teasing because of their appearance and short stature. Good RE or Personal and Social Education (PSE) policies need to examine the question of differences in race, religion, culture, appearance and disability. Children with Noonan Syndrome need a great deal of input and support to achieve in an increasingly complex society, but they are capable of achieving and ultimately giving back to the family and society that has supported them.

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