Learning Behaviour in Noonan Syndrome

Taken from presentation given at BDF Newlife Noonan Syndrome Information Day 21-11-03

Parents’ Questions:

- What do we know about NS child behaviour?
- What about NS and autism?

Learning difficulties

This is a subject of much debate, but generally speaking we learn many of our behaviours through:

- Example or models
- Social controls or patterns
- Responses to our actions
- Reinforcements – negative and positive
- Other influences – acquired or learned

Therefore, if the NS child is diagnosed as having a learning difficulty this may affect the way in which behaviours are learned and the pace at which they are learned.

Not all NS children have learning difficulties. Approximately one third of individuals have learning problems; others are of average, or even above average, intelligence.

What is behaviour?

There are several problems in discussing behaviour as a subject:

- Diagnosis variations – is the behaviour a result of the NS autism (ASD) or could it be a reaction to trauma, illness, treatment or hospitalisation?
- Difference in perception of normal behaviour - what is normal to one individual may be seen as abnormal by others because of difference in language, culture, habits or acceptance of the situation.
- Is the behaviour caused by the underlying condition (e.g. Noonan Syndrome) or is it reinforced by the response of others to that behaviour?

NS behaviour – What do we know?

- Some research has been carried out but much more is needed.
Past research has focused on a variety of factors, comparing different age ranges and the situation in different countries. Most of the information studied has been gathered by families. What behaviours have been reported?

- Stubbornness, cannot be reasoned with or persuaded
- Inflexibility, a need for a set routine
- Obsessions, negative or positive
- Poor eye contact
- Isolation; preferring his or her own company
- Inability to make friends or only able to make friends with younger children
- Immaturity, or appearing older than his/her age
- ‘Clingyness’, dependency, unresponsive or not liking to be touched
- Poor self-esteem
- Poor speech and/or understanding
- Clumsiness or poor body language
- Inability to understand the feelings of others
- Psychiatric disturbances (rare)

In some individuals there may be normal development and no distinctly different behaviours.

**Stubbornness – (cannot be reasoned with or persuaded)**

- Reported in most Noonan Syndrome research
- Commonly reported by parents
- May be linked to the child's need to ensure routine, safety or control or to overcome anxiety
- The child will often refuse to respond to 'bribery'
- There may be a 'verbal routine' which results in increased attention.
- In some children, may originate from an early 'feeding struggle'

**Stubbornness may have some positive benefits in the acquisition of academic progress or other skills**

**Inflexibility – (need for a set routine)**

- Often reported by parents
- Characterised by a need to follow elaborate plans or sequences (routines)
- Causes difficulties in family life
- Causes difficulties in relationships
- Timetables can often be used to create more orderly behaviour

**Obsessiveness – (negative or positive)**

He or she may be obsessive about:

- Themes, such as cars, music or sport
Words or phrases, which may be repeated or used wrongly
Physical actions, such as rocking or picking
A need for order, arrangement or sequence
Particular ideas
Relationships, manifesting in 'all or nothing'
Items such as toys, ornaments or collections

Obsessiveness may serve to create specialisation or extensive knowledge of certain interests

Poor eye contact
It is generally accepted that people/children with poor eye contact find it harder to make relationships, develop self esteem and communicate or interact with others. Poor eye contact gives the impression that the person is withdrawn. The individual may appear to be completely uninterested, with a 'don't care' attitude, often leading to misunderstanding. Aversion to one-to-one eye contact can induce the individual to look elsewhere, with fixed gaze, when being spoken to. Most people do not find this personally threatening.

Isolation, preferring his/her own company
There is a need to distinguish between:

A situation in which the individual is developing well and is obviously comfortable and content in his/her own company

A situation in which the individual is not developing well and appears to be negatively withdrawn, uncomfortable and unhappy.

An individual who is content with his/her own company may achieve a degree of positive self-reliance.

Inability to make friends or only makes friends with younger children
If the individual only makes friends or associates with younger children, it can reinforce immature behaviour or play
Interferes with the normal pattern of attaining maturity
Associated with poor self-esteem
Can lead to misunderstanding and false allegations
Can reinforce isolation or withdraw and emphasise other issues, such as height
Appearing 'too old' or 'too young' for his/her age
Behaving younger than his/her age – childish language and behaviour, inability to deal with bodily functions, slow development of skills, immature response to experiences, inability to detach or re-attach to others, immature movement, mimicking of others.
Behaving older than his/her age

– language and behaviour in excess of actual age, mimicking of individuals older than actual age, response to experiences in excess of actual.

'Clinginess' and dependency or unresponsiveness and aversion to being touched

The individual is ‘clingy’ and very physically dependent, with frequent touching or holding on to others. May lead to anxiety and withdrawal when separated from focus person and can result in misleading assumptions about age and ability.

The individual is unresponsive and does not like being touched, which reinforces their isolation. An aversion to physical contact may lead to an aggressive response or an increase in anxiety.

Poor self-esteem

Lack of self-esteem is often linked with poor body image in older children. It is recognised that those with poor body language and poor bodily control often have difficulty with relationships, which impacts on self-esteem. Depression and withdrawal, varying in severity and duration, is sometimes seen in older children, teenagers and adults. Some very useful and effective management techniques to overcome poor self-esteem have been established.

Poor speech/understanding

Many NS children require speech and language therapy. Some have problems with physical speech (mechanical pronunciation); others have difficulties with the use and comprehension of language. Some have a combination of both. Speech and language problems have an enormous effect on behaviour, level of anxiety, relationships and self-esteem.

OUTPUT – for those who cannot speak well or pronounce words clearly, being misunderstood cause great frustration. Because they do not sound like others, they also feel singled out as ‘different’. Feeling misunderstood and frustrated can result in an increase in negative behaviour.

INPUT – those who are unable to understand (comprehend) the words being spoken to them may not be able to respond or behave accordingly, reinforcing the feeling that they are ‘different’ from others. When tested, some NS children appear to understand more than they actually do.
Clumsiness and poor body language

It is generally accepted that those with poor control of the body can have difficulty with relationships and people may (often subconsciously) misunderstand or misread their movements. Bumping into things and clumsiness may be aggravated by, or associated with, lax muscles and/or skeletal anomalies. Some individuals tend to be accident-prone as a result of poor spatial skills. Clumsiness often improves with age and physical exercise or physiotherapy may be helpful.

Inability to understand the feelings of others

Some parents report that incidents, which severely upset them, seem to have little or no effect on their child. Even when experiencing a traumatic event, the child may not appear to be affected and may show no emotional response. The inability to empathise may have a negative effect throughout the life of the individual. The inability to understand the feelings of others is one of the features most strongly associated with autism (ASD). This characteristic is very difficult to assess correctly without professional skills and experience.

Psychiatric disturbances (rare)

Significant psychiatric disturbances do not appear to be more common or less common than in the general population. If psychiatric disturbances do occur they may be harder to diagnose and treat, due to condition overlap.

NS behavioural profile

NS does not have a specifically defined known, or formally recognised, behavioural phenotype (clinical picture), composed of a specific set of behaviours present in all or the majority of children (although some researchers may take a different view on this) however the NS behaviours are such that they are probably best described as a ‘behavioural profile’ for NS. Some individuals will have a few aspects of unusual or difficult behaviour. Other individuals will demonstrate several unusual or difficult behaviours. Many individuals will have no perceivable difference in their behaviour, apart from the normal variations found in the general population.

What else can affect behaviour?

If not handled well, the ordinary ‘difficult’ life experiences, such as bereavement, loss of a parent through separation, injury, bullying etc, can have a negative effect on the behaviour of any child. If the child has NS,
dealing with such situations may require more attention to the details, explanation and recovery time than with an unaffected child.

What else can affect behaviour? – Illness

It is generally recognised that medical intervention, treatment, therapy or surgery, especially if prolonged, dramatic or frequently repeated, may have relevance to negative behaviours in some children (but not all). This should not affect the decision to go ahead with treatments, but parents need to be aware that the child may need a greater input if they show an unusually dramatic or anxious response to the experience. Of course, many NS children have long-term experience of such interventions, treatment and surgery.

What about autism?

The classic symptoms of autism are not usually seen in NS. However, over the past few years many NS children have been diagnosed as having an ASD (Autistic Spectrum Disorder), in addition to NS.

Is this a transient diagnosing trend or NS reality? – The jury is still out.

Triad of impairment

An ASD has a 'triad of impairment' affecting:

- Social interaction
- Communication
- Imagination

These features are often seen in combination with a repetitive and stereotyped pattern of activities.

The word 'spectrum' (simply defined) refers to a scale which lists the extent of the features, ranging from severe impairment and loss of ability at one end, to the higher-performing end of the spectrum (as in Asperger Syndrome).

ASD symptoms

There is certainly some overlap in the symptoms and features of autism and those described in NS. Of the individual 13 points discussed here, 11 are reported also to exist in ASD.
NS & ASD overlap – ‘spot the *’

- Stubbornness, cannot be reasoned with or persuaded*
- Inflexibility, need for a set routine*
- Obsessiveness, negative or positive*
- Poor eye contact *
- Isolation, preferring his/her own company *
- Inability to make friends or only able to make friends with younger children *
- Immaturity, or appears too old for his/her age
- ‘Clinginess’, dependency, unresponsiveness, aversion to being touched *
- Poor self-esteem *
- Poor speech and/or understanding *
- Clumsiness and/or poor body language
- Psychiatric disturbances (rare) *

Understanding overlap

Example:

This has got:

- a hairy body, four legs and paws
- a wet nose and whiskers
- it has a tail
- it can often run fast and jump
- it is considered to be a pet
- it needs feeding

What is it?

It could be one of many things but most people would choose a cat or a dog. But, despite their long list of shared characteristics, they are very different creatures. So casual commonality isn’t enough. It is essential to have proper assessment by professionals who are experienced in dealing with complex overlapping NS disorders.

ASD diagnosis may help

In a world of resource restrictions and complex conditions, some parents have reported an improvement in provision when the ASD diagnosis is ‘attached’ to their NS child. This is probably because the educational and behavioural management of ASD is significantly better understood and provided for than NS. It may be that the perimeters of what is now called ASD, have become less rigid and the term is now used to encompass a wider range of special...
needs in children. If this applies to your child then you may receive improved services, to the child's long-term benefit.

However Autistic Spectrum Disorder is a significant diagnosis and, therefore, should not be lightly 'attached' to an individual by those without professional experience.

Diagnosis and Reasons

Diagnosis: Referral to an experienced specialist via your GP, hospital consultant or school psychology services.

Reasons: Professor Stephen von Tetzchner, in his NS book reports: "Autistic traits are often found in children with language difficulties and it is therefore natural that they are seen in children with Noonan Syndrome."

What now?

If this has prompted you to reassess some aspects of your child's behaviour, please seek professional help and guidance. Some families in the past have used the ABC method to help them identify and explain to others what is happening. Our nurses can give you more information on this. Don't forget that it helps to talk, so do call our helpline anytime, on 08700 70 70 20.

The beginning

Hopefully, this will be just the beginning of more research. We are hoping to launch further investigation into NS behaviour and autism, when funds and availability of researchers allow.

Remember – while most NS children share symptoms, appearance and particular behaviours, they are all very special individuals in their own right. Like all children, they should be treasured, supported and encouraged to develop their full potential – whatever that may be.