

Self-injurious behaviour in Cornelia de Lange Syndrome

A Guide for Parents and Carers

by

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Prof. Chris Oliver
University of Birmingham
12 June, 2003

Foreword

Parents of children with intellectual disabilities face a hard task in life. Of course, like most hard tasks, the experience is joyful at times - raising children who happen to have intellectual disabilities can be extremely rewarding. Nevertheless, families may sometimes feel isolated, particularly if they have a son or daughter with an unusual disorder, like Cornelia de Lange Syndrome. Even when they visit 'experts' in intellectual disabilities, such as paediatricians or psychiatrists or psychologists, families may find that they know more about their own child's syndrome than the 'expert' does. That can leave families feeling frustrated and unsure where to turn for advice.

Families need sound advice. There is plenty of homespun advice to be found but what families need is information about the true facts and advice about what will really help their child. This is especially the case when their son or daughter is engaging in a challenging behaviour, such as self-injury. Research has shown that having to cope with challenging behaviour is a major stressor for families. Self-injury is probably one of the hardest behaviours to deal with. It is extremely difficult for loving families to keep a cool head, in the face of self-injury, because their instinct is to protect the child and give him or her what s/he wants, so as to get the self-injury to stop. And yet research shows this can sometimes

be an unwise strategy, because it may teach the child to use self-injury like a communication device.

This book is written especially for families who are seeking to find the best information there is on self-injury in Cornelia de Lange Syndrome. It will also be helpful for parents whose children show self-injury but do not have Cornelia de Lange Syndrome. The research team involved are very well respected and they have worked closely with the Cornelia de Lange Syndrome Foundation over a period of years, collecting information and conducting research. The work is world class. Let's hope the so-called 'experts' read it too.

And, lastly, a word of encouragement for the parents, who may be worrying about the effects of all this on the rest of their family. Research evidence suggests that the experience of having a child with disabilities often helps bind families together (so that divorce rates, for example, are lower in such families). Moreover, although parents often worry about the effects on other children in the family, research evidence shows that siblings survive well and are more likely to enter the caring professions, than children who do not have siblings with disabilities. So enjoy this book and keep going, it will be worth it.

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Chapter 1: Introduction

1.1: Who this book is for

This book has been written for parents and carers to help towards an understanding of self-injurious behaviour when it is shown by a child or adult with Cornelia de Lange Syndrome. The book is not meant to be a comprehensive manual for assessments and interventions but is a guide to causes of self-injury and the strategies and resources that are needed in order to reduce this distressing behaviour. By writing this book we hope that parents and carers will become informed about the causes of self-injurious behaviour and active contributors to the assessment and treatment process and so be in a position to help professionals who may have limited experience of Cornelia de Lange Syndrome.

1.2: Why we have written this book

There are a number of reasons why we felt it was necessary to write this book. The main one is that although there has been an increase in the amount of information about behaviour disorders and syndromes, it is not easy to obtain. Most of the information about both self-injurious behaviour and Cornelia de Lange Syndrome is generally inaccessible to carers and parents as well as many professionals. Research articles are scattered throughout academic journals published over decades and periodically they need to be brought together, reviewed and interpreted. Many of the books that have been written on self-injurious behaviour are expensive to purchase and are often found only in the libraries of universities. On the few occasions that the information on Cornelia de Lange Syndrome and self-

injurious behaviour has been collated there is a lack of integration. Texts on self-injurious behaviour rarely refer to Cornelia de Lange Syndrome, except in passing, and vice versa. Separation of the two topics does not make the task of understanding and treating self-injurious behaviour in Cornelia de Lange Syndrome any easier.

It is estimated that there is a ten year delay between research being conducted and the findings being put into practice. That is too long. Additionally, the amount of research funding that is available to study self-injurious behaviour in Cornelia de Lange Syndrome is extremely limited in the UK and this situation appears unlikely to change. In combination, these factors mean that research will not have an immediate and significant impact on the lives of children and adults with Cornelia de Lange Syndrome unless a short cut can be found. Whilst it might be preferable to wait for research to uncover all of the reasons for self-injurious behaviour in Cornelia de Lange Syndrome this will take a long time. Many children and adults with Cornelia de Lange Syndrome cannot wait for this research to be conducted so we have to use what we have and try to fill the gaps in knowledge with what we know from related research. The second reason for writing this book is that many parents and carers want to act now.

1.3: Self-injury in children and adults who do not have Cornelia de Lange Syndrome and other behaviour problems

In addition to parents and carers of children and adults with Cornelia de Lange Syndrome we hope that some parts of the book will be useful

to any parent or carer of a child or adult who has an intellectual disability. In particular the information in chapters 3, 5, 6 and 7 is not specific to Cornelia de Lange Syndrome. Also, we hope the idea of trying to understand how syndrome specific physical and psychological features and theories of the causes of self-injurious behaviour might be integrated, might be of interest to parents and carers of other individuals who show self-injurious behaviour and have a genetically determined syndrome, such as Cri du Chat or Smith-Magenis syndromes in which self-injury is common. Finally, most aspects of the psychological theories of why self-injury occurs are not necessarily specific to self-injurious behaviour and can be generalized to other types of challenging behaviour such as aggression, shouting and screaming and destroying the environment. This information is presented in Chapter 5.

1.4: The sources of information for this book

The information and perspectives that are presented in this book come from a number of different sources. Over the last 20 years we have conducted a number of research projects into the prevalence, causes and treatment of self-injurious behaviour in children and adults who have an intellectual disability. Whilst much of this research has not been specific to Cornelia de Lange Syndrome many of the general principles are clearly applicable to all individuals who have an intellectual disability including those with Cornelia de Lange Syndrome. Some of the findings come from research conducted by Phillipa Hyman, a clinical psychology postgraduate at the University of Birmingham between 1996 and 1999. Much of the information in chapters 2 and 4 comes from two postgraduate theses that were written by Kate Arron and Jenny Sloneem whilst studying at the University of Birmingham in the UK between 1999 and 2002 and working on a project that was funded by the Community Fund via the Cornelia de Lange Syndrome Foundation (UK and Ireland) and the Medical Research Council. The projects conducted during this time,

and subsequently the work of Joanna Moss and Jane Petty, were supervised by Scott Hall and Chris Oliver.

These projects have supplied us with research evidence for the perspective that we propose throughout the text. However, this is not always the richest source of information. In addition to reviewing literature and conducting research, an extremely important source of information has been our clinical work and attendance at Cornelia de Lange Syndrome conferences in the UK, Ireland, Italy and the USA. At these conferences we were very fortunate to be able to speak to a number of parents and carers who described self-injurious behaviour of children and adults with Cornelia de Lange Syndrome in some detail and allowed us to gain insight into some of the reasons that the self-injury might occur. We hope that this combination of sources of information has allowed us to cover most of the areas that parents and carers want to know more about.

1.5: Parents and professionals

We noted at the outset that this book is primarily aimed at parents and carers. The purpose of the book is to guide parents and carers through the process of understanding self-injurious behaviour and being actively involved in conducting assessments and interventions. It can be difficult to find the right kind of professional help and many parents may want to try things for themselves. Some tasks are easier than others and the severity of the self-injury and the personal resources of time, energy and support will often be the determining factors in deciding how much and what to take on. At any of the stages when parents and carers are tackling self-injurious behaviour it is important to at least seek advice if not help. We have tried to indicate those professionals who might be able to offer the most appropriate advice at each stage. Finally, throughout the book we have provided references to scientific journal articles, books and book chapters so that parents and carers might look at the evidence and professionals might

seek more detailed and technical information. At the end of the book we have also provided a bibliography for further reading on some topics.

1.6: How to use this book

The book has been structured to guide parents and carers through the process of understanding, assessing and intervening. There are four main sections to the book:

- ❖ Background information about relevant features of Cornelia de Lange Syndrome.
- ❖ Information about self-injurious behaviour in people with intellectual disability and what is known about various aspects self-injurious behaviour in children and adults with Cornelia de Lange Syndrome, including potential causes.
- ❖ Descriptions of assessment strategies that will help parents and carers to relate the general information presented in the causes section to the person they know and to build a model of the possible causes of self-injurious behaviour so that an intervention strategy can be developed.
- ❖ Information on the different types of intervention strategy that can be used given the model of possible causes that has been developed for the individual. In this section the emphasis is on both developing rational interventions and systematic evaluation of how effective they are.

We strongly recommend that those using this book do not skip the early sections on background and assessment as we believe these are critical to increasing the chances that an intervention will be successful.

1.7: What can be achieved

It must be stated at the outset that there are no quick, easy or 'one size fits all' solutions to the problem of self-injurious behaviour in children and adults with Cornelia de Lange Syndrome.

Self-injury is not like a disease with a single cause and clear pathway to treatment. In the majority of people with intellectual disabilities who show self-injurious behaviour it has become clear that the causes vary considerably between individuals and that for any individual there may be a number of factors that contribute to self-injurious behaviour and these may well change over time. We believe the strategy to adopt is one of trying to understand as much of the self-injurious behaviour as possible so that a rational intervention can be implemented for that part which we understand. When we cannot identify a particular cause then we may need to rely on using a systematic approach to implement interventions that have been shown to work for others. Whilst this situation is less than desirable it is certainly better than doing nothing.

Some interventions are, without doubt, demanding of resources. This is particularly true of those interventions that involve changes to the way self-injurious behaviour is managed and increasing peoples' adaptive behaviours. It is quite likely that access to these resources will be difficult as they are not necessarily well supported within the National Health Service. However, our experience is that parents who lobby their local services and are persistent in seeking services are generally more successful than those who take 'no' for an answer. Additionally, even if the ideal intervention cannot be put into place because there is a lack of resources then some compromise may be necessary. Once again we believe that doing something is better than doing nothing.

It is also important to note that although there is very little research on the persistence of self-injurious behaviour in children and adults with Cornelia de Lange Syndrome over time we have collected some evidence that shows that the behaviour does appear to be very persistent. It is likely therefore that a long-term approach to self injurious behaviour is the most appropriate. This should not be cause for pessimism as for many people self-injurious behaviour may persist at a very low and unproblematic level. However, for

some people the behaviour may come and go and it is important to come back to this book on a number of occasions rather than using it as a one-off. Even though we know there is evidence that self-injurious behaviour may persist over time our experience is that it is possible to reduce the behaviour. Our advice therefore is do not give up, keep trying different strategies and through a process of trial and error you will arrive at an approach that will be the most beneficial for the person who shows self-injurious behaviour.

1.8: A comment on Behavioural Phenotypes

Before going any further we want to introduce the idea of a 'Behavioural Phenotype' and give our perspectives on the relevance of this idea to people with Cornelia de Lange Syndrome. 'Behavioural Phenotype' is the term used to describe a strong association between specific behaviours and a particular genetic syndrome. It is often assumed that because of this association, the behaviours arise from the genetic make-up of individuals with the syndrome. Much research has been conducted in this area and published reports have described associations that are believed to exist. Commonly accepted examples include self-injurious behaviour in Lesch-Nyhan Syndrome, hyperphagia (over-eating) in Prader-Willi Syndrome and stereotyped, repetitive hand movements in Rett Syndrome¹.

It has been suggested in the research literature that a behavioural phenotype exists for Cornelia de Lange Syndrome and that one of the behaviours that exists as part of the phenotype, is self-injurious behaviour^{2,3}. This assumption can be traced to William Nyhan and his presidential address to the Society for Pediatric Research in 1972⁴. At this time Dr. Nyhan was studying what became known as Lesch-Nyhan Syndrome in which self-injurious behaviour is almost always seen and is frequently very severe. This led Nyhan to suggest that the behaviour was due to a biological disorder in Lesch-Nyhan Syndrome. It would seem that Nyhan then made the obser-

vation that severe self-injurious behaviour was seen in Cornelia de Lange Syndrome and consequently suggested that there might also be a single biological cause. Since this time an assumption has developed that self-injurious behaviour might be an unchangeable feature of Cornelia de Lange Syndrome.

Since the early observations of Nyhan it has become possible to see why this idea might have been put forward. Firstly, as Nyhan pointed out, because he had an interest in self-injurious behaviour it is likely that he was referred mainly children and adults with Cornelia de Lange Syndrome who showed self-injurious behaviour. Consequently, he did not see all children and adults with Cornelia de Lange Syndrome and so did not see those who did not engage in the behaviour. Additionally, self-injurious behaviour is seen in some children and adults who have an intellectual disability, but who are not diagnosed with any specific syndrome, thus self-injury is not necessarily associated with any particular genetic syndrome. As research has progressed in this area, it has become clear that whilst self-injurious behaviour is common in Cornelia de Lange Syndrome, it certainly is not an inevitable feature of the syndrome. This is important as it suggests that as there is no one-to-one relationship between self-injurious behaviour and Cornelia de Lange Syndrome, then the situation is very different to that for Lesch-Nyhan Syndrome, where self-injury is believed to be genetically determined and consequently inevitable.

More recent views have taken a slightly different perspective to behavioural phenotypes than that discussed by Dr. Nyhan, reflecting the many possible mechanisms within biology and the environment that may shape and effect how we behave^{5,6}. As mentioned above, we believe that a behavioural phenotype is a characteristic pattern of behaviours that are strongly associated with a specific syndrome. However, whereas some people suggest that a biological mechanism underpins the behaviour, we make no assumptions as to the mechanism causing the association. Instead, it is sometimes useful sim-

ply to think of a behavioural phenotype as the increased likelihood that a person with a genetic syndrome will exhibit certain behavioural or developmental features when compared to a person without the syndrome⁵. Thus, behaviours seen commonly in syndrome groups may not necessarily be determined by the genetic make-up of the syndrome, but may be influenced by other factors. Box 1.1 (facing page) shows how, using this perspective, a number of behavioural phenotypes have been described in relation to specific genetic syndromes.

A greater insight into the associations between genetic syndromes and behaviour would help toward an understanding of syndrome-behaviour relationships and the reasons why the associations exist. Understanding in this area would also help to guide treatment and education. By these means, standard tasks and lessons may be tailored to meet the needs of individuals with greater understanding of their intellectual and behavioural strengths and needs. The improvement in education and development, together with the possible reduction in problem behaviours, which are often the reason for failed care and social exclusion, has the potential to increase the quality of life in many individuals.

Although self-injurious behaviour is common in children and adults with Cornelia de Lange Syndrome, there is certainly no need to think that the behaviour cannot be reduced. It can. Additionally, some of the causes of self-injurious behaviour in Cornelia de Lange Syndrome are likely to be similar to those seen in all children and adults who have intellectual disability. If we only think about self-injurious behaviour in Cornelia de Lange Syndrome we run the risk of dismissing useful ideas and information that come from a much broader approach.

It may well prove to be the case that there is a Behavioural Phenotype for Cornelia de Lange Syndrome i.e. that some cognitive and behavioural features do associate to the syndrome. However, these cognitive and behavioural features do not define children and adults with

Cornelia de Lange Syndrome. Children and adults who have Cornelia de Lange Syndrome are people first and have more in common with other children and adults than they have differences. The main task for us is to try and understand why particular features of Cornelia de Lange Syndrome might be associated with self-injurious behaviour and also to take information from the broader literature on all people with intellectual disabilities and work out the extent to which this is applicable to people with Cornelia de Lange Syndrome.

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Box 1.1: Genetic syndromes and their reported Behavioural Phenotype.

Cornelia de Lange syndrome

Prevalence: 1 in 50,000⁸.
Genetic Anomaly: Chromosome 3, unknown location^{9,10}.
Physical Features: Low birth weight, delayed growth, thin down-turned lips, long eyelashes, confluent eyebrows, limb abnormalities^{11,12}.
Behavioural Phenotype: Intellectual disability, poor communication, self-injury, aggression, hyperactivity, compulsive behaviour, stereotyped behaviour, and “autistic like” behaviour^{13, 14, 15, 16}.

Angelman syndrome

Prevalence: 1 in 12-25,000^{17, 18}.
Genetic Anomaly: Chromosome 15, location q11-q13¹⁹.
Physical Features: Low-birth weight, jerky movements, long face, prominent jaw, wide mouth, widely spaced teeth, protruding tongue, deep set eyes²⁰.
Behavioural Phenotype: Severe intellectual disability, happy demeanour, mouthing objects, sleeping problems, feeding problems, hyperactivity, inattention, and hand flapping^{21, 22, 23, 24}

Cri du Chat syndrome

Prevalence: 1 in 50,000²⁰
Genetic Anomaly: Chromosome 5, location 5p1524.
Physical Features: Cat-like cry, a round face with epicanthal folds, down-slanting palpebral fissures, low-set malformed ears, growth delay.²⁶
Behavioural Phenotype: Severe intellectual disabilities, motor and language delay, hyperactivity, hypersensitivity to sensory stimuli, self-injurious behaviour and obsessional behaviour^{26, 27, 28}.

Lesch-Nyhan syndrome

Prevalence: 1 in 380,000²⁸.
Genetic Anomaly: Chromosome X, location q26-27, males only²⁹.
Physical Features: Delayed motor development, hypotonia, feeding difficulties, low height and weight, microcephaly, seizures, urinary infections³⁰.
Behavioural Phenotype: Intellectual disability, motor delay, choreoathetosis, cerebral palsy, “compulsive” self-injurious behaviour and physical aggression^{31, 32}.

Rett syndrome

Prevalence: 1 in 10-15000³³.
Genetic Anomaly: Chromosome X, location q28 females only³⁴.
Physical Features: Breathing dysfunction, ECG abnormalities, growth retardation, seizures, scoliosis, deceleration in head growth and loss of facial expression^{35, 36}.
Behavioural Phenotype: Normal development, followed by deterioration of skills and severe intellectual disability, autistic-type behaviour, repetitive hand movement and anxiety^{37, 38, 39}.

Prader-Willi syndrome

Prevalence: 1 in 16,000⁴⁰.
Genetic Anomaly: deficient expression of paternally expressed imprinted genes at 15q11-13, minimal region of deletion between loci D15S13 and D15S10⁴¹
Physical Features: Short stature with small hands and feet and scoliosis. Facial features include almond shaped eyes, thin upper lip, triangular shape mouth and a narrow nose⁴².
Behavioural Phenotype: Mild to moderate intellectual disability, hyperphagia tantrums, obsessive and compulsive behaviours^{42, 43, 44}.

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Chapter 2: Cornelia de Lange Syndrome

2.1: Introduction

Cornelia de Lange Syndrome is named after a Dutch paediatrician who described the syndrome in 1933. The disorder is occasionally referred to as Brachmann de Lange Syndrome after a German doctor who is now known to have described the syndrome in 1916.

Box 2.1: Cornelia de Lange, Professor of paediatrics.

Cornelia de Lange was a Professor of Paediatrics who specialised in the field of micropathology of the brain. In 1933 Cornelia de Lange described two children with the syndrome. Despite the fact that these two girls were not related, their remarkable facial similarities led to both the nursing staff and Cornelia de Lange herself to confuse them as being the same person. In 1941 having described a third patient with the disorder, she presented an account of the syndrome at a meeting of the Neurological society of Amsterdam. She named the syndrome 'typus degenerativus Amstelodamensis'. It has since then taken her name, but may also be referred to as Brachmann de Lange syndrome in acknowledgement of Brachmann, a German doctor who is now known to have written the first description of the syndrome in 1916.

At present the cause of Cornelia de Lange Syndrome is unknown. It is believed to be caused by a genetic anomaly on the long arm of chromosome 3 although so far, research into the specific location of the gene abnormality has

been inconclusive^{1,2,3}. It is thought however, that the anomaly is probably a de novo (new and not passed on) mutation and consequently the syndrome does not tend to run in families¹. Without a genetic marker, diagnosis of Cornelia de Lange Syndrome relies on the recognition of the characteristics associated with the syndrome⁴.

A number of physical, medical and psychological features are thought to be associated with Cornelia de Lange Syndrome. In this chapter we aim to provide an overview of the main issues, focusing on those features that might be relevant to self-injury. More detailed information can be obtained from the Cornelia de Lange Syndrome website.

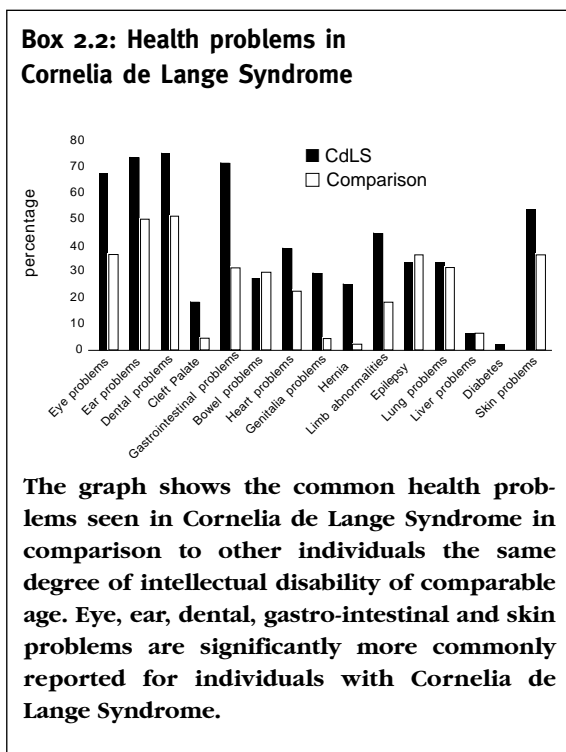
2.2: Medical and physical features of Cornelia de Lange Syndrome relevant to self-injury

The physical characteristics of Cornelia de Lange Syndrome have been described by a number of researchers and clinicians^{4, 5, 6, 7, 8, 9}. The most prominent characteristics include:

- ❖ Low birth weight.
- ❖ Delayed growth.
- ❖ Distinctive facial features (prominent philtrum (this is between the nose and the upper lip), thin downturned lips, long eyelashes, confluent (joined) eyebrows)
- ❖ Excessive hair growth
- ❖ Small hands and feet with short digits.
- ❖ Severe limb abnormalities (present in 25-30%).

Many health and medical problems are also associated with Cornelia de Lange Syndrome. Box 2.2 shows how common some of these problems are in children and adults with

Cornelia de Lange Syndrome in comparison to other children and adults with intellectual disabilities of the same age and with the same degree of intellectual disability.



For the purpose of understanding self-injurious behaviour in Cornelia de Lange Syndrome, this chapter will attend more closely to those physical characteristics and health or medical problems that may be associated with discomfort or pain. This is because a number of these problems are potentially relevant to self-injurious behaviour. These are described below together with an indication of why they might be associated with self-injury.

Eye problems^{6, 10}. The primary eye problems associated with Cornelia de Lange Syndrome include the absence or reduction of tears, blocking of the tear duct and potential abnormalities of the eye that cause impaired vision. In many people who have intellectual disabilities discomfort, irritation or pain in the eye will lead to eye pressing, pok-

ing or rubbing. Additionally, if vision is impaired by damage to the eye but the optic tract (the nerve fibres that carry signals from the eye) is intact and the occipital lobe (this is the part of the brain that receives and interprets signals from the eye) is functioning normally, then this can lead to eye pressing or poking. The reason for this is that if no light can stimulate the nerves in the back of the eye then physical stimulation of these nerves by pressing or poking leads to bright flashes of light and light patterns. In the absence of any other stimulation this can be rewarding and cause the behaviour to occur again in the future (see Chapter 5). Another reason for pressing or poking the eye is that this can lead to the shape of the eye being temporarily changed and consequently focus is improved.

Ear problems^{5, 6, 10}. The most commonly reported ear problem in Cornelia de Lange Syndrome is chronic otitis media (middle ear infection). Approximately 50% of head-banging that is commonly seen in many children under the age of five, who do not have intellectual disabilities is associated with this problem³¹. There is no reason to think that children with Cornelia de Lange Syndrome should respond any differently to this painful condition than children without intellectual disabilities.

Bone and joint problems^{5, 12}. Although there has been limited research into bone and joint problems in Cornelia de Lange Syndrome the presence of scoliosis (curvature of the spine), difficulties with gait and restricted movement in certain joints such as the elbow have been reported. Abnormalities in the hip occur in approximately 5-10% of children and adults with the syndrome. Some parents also indicate that their children often 'pop' their hips, knees and other joints out of place. However, research on this has not been carried out and at the moment these reports are anecdotal. It is not clear what sensation is gained from this behaviour but it is thought that perhaps it might help to relieve pain or discomfort in the joints.

Gastro-intestinal disorders^{5, 6}. Gastro-intestinal problems are one of the most commonly reported problems in Cornelia de Lange Syndrome. Research suggests that as many as 71% of children with the syndrome have feeding difficulties that reflect these problems.

There are two main gastro-intestinal disorders that are common in children and adults who have Cornelia de Lange Syndrome. The first is gastroesophageal reflux. This is caused by the sphincter muscle at the top of the stomach not being sufficiently tight to keep the contents of the stomach from rising up into the oesophagus. The contents of the stomach are acidic causing painful burning sensations in the chest and throat when reflux occurs. The second problem is malrotation of the intestine. This is a serious and life-threatening condition that leads to considerable discomfort and requires immediate attention.

*Sinuses*¹². Sinusitis has been reported to be a common problem for individuals with Cornelia de Lange Syndrome. It is caused by inflammation of the sinuses causing pain and discomfort to the individual. Sinus problems may be related to the depressed nasal bridge that is characteristic of individuals with the syndrome.

Teeth^{5, 11}. There are two dental related medical problems that might be associated with self-injurious behaviour. The first is the crowding of teeth. This can lead to some discomfort as teeth erupt. The second problem is the thin layer of enamel on the teeth. This may mean that dental cavities are more likely to occur and that there is increased sensitivity to touch and temperature in the teeth. This is potentially relevant to gastroesophageal reflux as constant contact between the acidic fluid from the stomach and the already thin enamel on the teeth can hasten the development of cavities.

In addition to these two specific problems that are related to Cornelia de Lange Syndrome it should be noted that there will in any event be some physical discomfort around the time of

tooth eruption just as there would for any child.

*Peripheral sensory neuropathy*³². Dr. Tonie Kline has suggested that here is some evidence that children and adults who have Cornelia de Lange Syndrome might have a peripheral sensory neuropathy. This means that the peripheral nervous system, that part of the nervous system outside of the brain and spinal cord that is associated with pain and sensation amongst other things, might not be sending the right signals to the brain. Given that for many children and adults the upper limbs do not develop properly it would not be surprising if the peripheral nervous system in the arms had also not developed normally. Even if the hands and arms are well-formed it is entirely possible that a peripheral sensory neuropathy is still present.

This disorder can have two important effects that might be relevant to self-injury. First, pain may not be experienced in the 'normal' way. Painful stimuli may not be experienced as painful. Secondly, people may experience unusual sensations in their hands and arms. These sensations are described by some people as pins and needles (dysaesthesia) or a mild burning sensation.

2.3: Psychological factors relevant to self-injury

As discussed in Chapter 1, a number of cognitive and behavioural features, i.e. a behavioural phenotype, may be associated with Cornelia de Lange Syndrome. In order to help us to understand self-injurious behaviour in Cornelia de Lange Syndrome, it is important to consider the cognitive and behavioural features that might be related to the syndrome. The specific features of the behavioural phenotype, including those that are relevant to self-injury are described here, together with a brief indication of why they might be important to consider in relation to self-injury. Chapter 5 provides further insight into why and how these features might be associated with self-injurious behaviour in Cornelia de Lange Syndrome.

Intellectual disability. One of the main psychological factors that is relevant to self-injurious behaviour in Cornelia de Lange Syndrome is the associated degree of intellectual disability. Research has indicated that people with Cornelia de Lange Syndrome have an intellectual disability ranging from mild to profound degree of disability, with the majority of people having a severe disability^{5, 7, 13, 14, 15, 16, 17}. In our research project we found that 45.6 % of people with Cornelia de Lange Syndrome had a profound degree of intellectual disability and 30.43% had a severe degree of intellectual disability when we used a standardised measure of adaptive behaviour (see Box 2.3). Presence of self-injury is related to intellectual disability and research has suggested that individuals with a greater degree of disability are more likely to display self-injury^{18, 19} (see Chapter 3).

Box 2.3 Degree of intellectual disability.

The table below shows the percentage of individuals in each category of degree of intellectual disability in our study of 54 people with Cornelia de Lange Syndrome. Note that the majority of individuals with Cornelia de Lange Syndrome have either profound or severe intellectual disabilities.

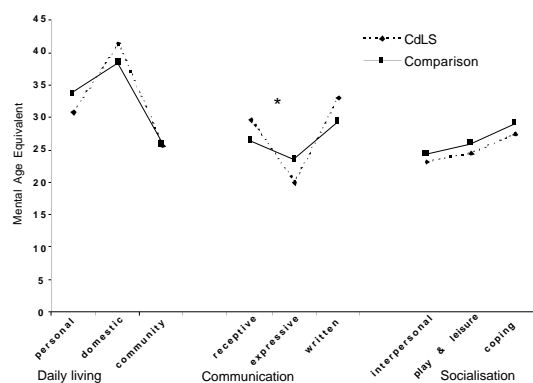
Percentage of individuals in each category of degree of intellectual disability

	Profound	Severe	Moderate	Mild	Borderline
CdLS	45.65	30.43	15.22	8.70	0.0

Communication. Past research indicates that children and adults with Cornelia de Lange Syndrome have communication deficits with around 30-85% of individuals showing no verbal communication skills^{15, 16, 18, 20, 21, 21, 23}. It is thought that the main problem related to communication is a deficit in expressive communication (see Box 2.4). Although children and adults with Cornelia de Lange Syndrome may have difficulties with expressive communication, research has indicated that individuals with Cornelia de Lange

Syndrome use a number of non-verbal strategies to communicate including, for example, approach, touch and pushing a person’s hand away²². Whilst this problem of poor expressive communication is not unique to Cornelia de Lange Syndrome (it is also seen in both Cri du Chat and Angelman Syndrome for example) it does have a particular relevance to self injurious behaviour (see Chapters 3 and 5).

Box 2.4: Adaptive behaviour in Cornelia de Lange Syndrome

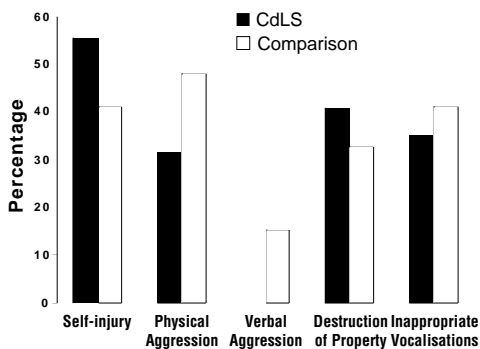


* = This indicates the presence of a significant difference between the Cornelia de Lange Syndrome group and the comparison group. The graph shows that individuals with Cornelia de Lange Syndrome have a particular deficit in expressive communication (ability to express information to others) compared to other individuals with intellectual disabilities. The graph also indicates that individuals with Cornelia de Lange Syndrome have significantly better receptive communication (ability to understand incoming information from others) than this comparison group.

Self-injury and aggression. As mentioned in Chapter 1, self-injurious behaviour has been suggested to be associated with Cornelia de Lange Syndrome^{14, 20, 22, 24, 25, 26, 27, 28}. Initially it was reported that self-injury was a significant feature of the syndrome. However, our research has shown that the association is not as strong as was at first thought. This is discussed in more detail in chap-

ter 4. In addition to self-injurious behaviour, children and adults with Cornelia de Lange Syndrome can show other problem behaviours including physical and verbal aggression towards others and destruction of the environment (see Box 2.5). Interestingly, whilst there is no significant difference between the percentage of children and adults with Cornelia de Lange Syndrome who show self-injurious behaviour or destruction of the environment, children and adults with Cornelia de Lange Syndrome show significantly less aggression than children and adults who have the same degree of intellectual disability.

Box 2.5: Prevalence of self-injurious behaviour and other behaviours in Cornelia de Lange Syndrome

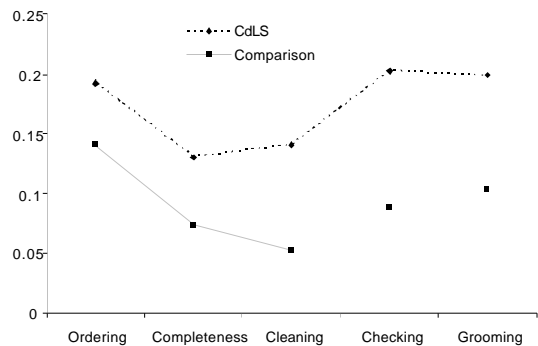


The graph shows that whilst individuals with Cornelia de Lange Syndrome show high rates of self-injurious behaviour, other behaviours such as physical aggression and destruction of property are also common. However, it is important to note that children and adults with Cornelia de Lange Syndrome show significantly less physical aggression than others with intellectual disability.

Compulsive behaviour. Our recent research shows that children and adults with Cornelia de Lange Syndrome show significantly more compulsive behaviours than children and adults with same degree of intellectual disability (see Box 2.6). This is potentially important and relevant to self-injurious behaviour because it has been suggested that compulsive behaviours are related to

severe self-injury that the person finds difficult to control³³. A number of studies have suggested that self-injury in people with Cornelia de Lange Syndrome has a compulsive quality and is associated with compulsive behaviour^{23, 27, 28}. (This is discussed in detail in Chapter 5).

Box 2.6: Compulsive behaviour in Cornelia de Lange Syndrome



Compulsive behaviours have been defined as ‘repetitive, intentional behaviours that appear to follow certain rules’. According to some researchers as many as 87.5% of individuals with Cornelia de Lange Syndrome show at least one form of compulsive behaviour²³. The graph below shows that individuals with Cornelia de Lange Syndrome show significantly more compulsive behaviours than the comparison group. In particular, checking, ordering and ‘grooming’ are among the most common compulsive behaviours in individuals with the syndrome.

Hyperactivity. In the past it has been suggested that hyperactivity is a feature of Cornelia de Lange Syndrome^{20, 29}. In our recent research we could find no evidence that hyperactivity was more common in Cornelia de Lange Syndrome than in children and adults with the same degree of intellectual disability. This does not mean that it does not occur, only that it is no more common than we would expect by chance.

Autism. Autism and autistic type behaviour has also been reported to be associated with Cornelia

de Lange Syndrome^{20, 25}. However, in our survey of people with Cornelia de Lange Syndrome we found different results for autism depending on the measure that we used. One measure (Gilliam Autism Rating Scale) showed no difference between the two groups whilst a second measure (Childhood Autism Rating Scale) showed a statistically significant difference between the two groups with autism being more common in children and adults with Cornelia de Lange Syndrome (see Box 2.7). It is important to clarify this issue as autism has been identified as a risk marker for the development of self-injurious behaviour³⁰ (see Chapter 3).

At present it is unclear whether autism is associated with Cornelia de Lange Syndrome and further research is needed. For both hyperactivity and autism if there is no association with Cornelia de Lange Syndrome it does not necessarily mean that these do not occur, it only means that they are no more likely to occur in children and adults with Cornelia de Lange Syndrome than in people who have the same degree of intellectual disability.

Stereotyped behaviour. Finally, the presence of stereotyped behaviours also warrants mention, since there is some suggestion that there is a link

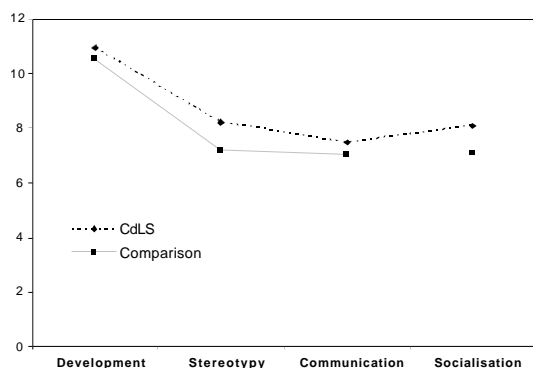
Box 2.7: Autism in Cornelia de Lange Syndrome.

There are three core deficits that are currently used to diagnose the presence of autism:

- 1. Impairment in reciprocal social interaction.**
- 2. Impairment in verbal and non-verbal communication.**
- 3. Presence of restricted and repetitive behaviours.**

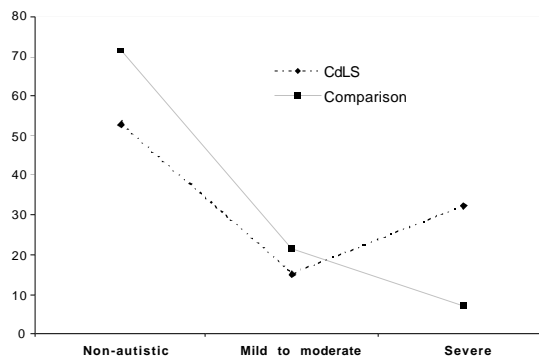
The diagnosis of autism will only be made if all three of these impairments are present.

Two instruments that measure the presence of these three impairments are the Gilliam Autism Rating Scale and the Childhood Autism Rating Scale.



Graph 1: Presence of autism according to the Gilliam Autism Rating Scale

According to this graph individuals with Cornelia de Lange Syndrome do not show significantly increased levels of impairment on communication, socialisation and the presence of repetitive behaviour compared to other individuals with intellectual disabilities. According to this, 'levels of autism' are no different between the two groups.



Graph 2: Presence of autism according to the Childhood Autism Rating Scale

In contrast to Graph 1, this graph indicates that individuals with Cornelia de Lange Syndrome show significantly increased occurrence of severe autism compared to other individuals with intellectual disabilities.

More research is needed to establish the relationship between autism and Cornelia de Lange Syndrome.

between these behaviours and self-injurious behaviour. Stereotyped behaviours can be defined as repetitive, apparently purposeless body movements or use of objects e.g. body rocking or spinning objects. There has been little research into stereotyped behaviour in Cornelia de Lange Syndrome, however studies that have been carried out suggest that between 57% - 78% of individuals with the syndrome show stereotyped behaviour^{22, 24, 25}. This is not thought to be significantly different from other individuals with intellectual disabilities and the most common forms of this behaviour in Cornelia de Lange Syndrome reported in previous studies include body rocking, bizarre body positioning and object spinning^{22, 25}.

In our recent observational study of children and adults with Cornelia de Lange Syndrome, we found that significantly more individuals with Cornelia de Lange Syndrome showed distinct rigid J or C-shaped hand posturing than individuals with the same intellectual disability but without Cornelia de Lange Syndrome.

2.4: Summary

Whilst there are many ways in which children and adults with Cornelia de Lange Syndrome are no different from other individuals both with and without intellectual disabilities, there are some ways in which they are significantly different. The most important differences with regard to self-injurious behaviour are probably the medical issues that can be associated with irritation, discomfort and pain, and the psychological issues such as the degree of disability that is associated with Cornelia de Lange Syndrome, the presence of compulsive behaviours, the possible association with autism and more specific characteristics such as poor expressive communication and the presence of hand posturing.

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Chapter 3: Self-injurious behaviour in people with intellectual disability

3.1: What is self-injurious behaviour?

Self-injurious behaviour can be seen in a number of different populations and in both children and adults. People who are depressed may attempt suicide and a very small minority of people experiencing psychosis may injure themselves in response to the auditory hallucinations¹. Self-injury is also seen in penal institutions^{2,40} (see Box 3.1) and delicate cutting with glass and blades is sometimes shown by young women in association with borderline personality disorder. self-injurious behaviour in people with intellectual disability tends to be a different than that shown by these populations. It tends to be cruder in terms of the act itself (e.g. self-biting or head banging), does not normally involve the use of objects and is not an attempt at suicide.

Box 3.1 Self-injury in penal institutions

In her book *Gulag: A history*,⁴⁰ Anne Applebaum describes the practice of *samorub* (self-mutilation) in the notorious Soviet concentration camps.

“Some of the methods were crude. Criminals in particular were famous for simply cutting off their three middle fingers with an axe, so they could no longer cut tress or hold a wheelbarrow in the mines. Others cut off a foot, or a hand, or rubbed acid into their eyes. Still others, upon departing for work, wrapped a wet rag around one foot: in the evening they returned with third degree frost-bite.”

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In the research literature a number of different terms have been used to describe these behaviours in people with intellectual disability including self-mutilation, automutilation, auto-plexy, self-harm and self-abuse. The term self-injurious behaviour is preferred because it can include behaviours which do not necessarily result in mutilation, for example face slapping, and is more specific than terms such as self-harm and self-abuse.

Inevitably there is some debate as to what might be included under the term self-injurious behaviour and consequently some definitions have been developed. Perhaps the most useful is that given by Tate and Baroff³: “Self-injurious behaviour does not imply an attempt to destroy, nor does it suggest aggression; it simply means a behaviour which produces physical injury to the individual’s own body... Common types of self-injurious behaviour are forceful head-banging, face slapping, punching the face and head and scratching and biting one’s own body”. However, even when this definition is applied to exclude less severe behaviours it is clear that some behaviours that should be included, such as face slapping that leads to reddening of the face but not necessarily tissue damage, are excluded. However, the definition does have the benefit of not including more mild behaviours that are not really injurious.

Other terms that are associated with self-injurious behaviour tend to refer to different aspects of behaviour. The term “challenging behaviour” has replaced those of the “problem behaviour” and “behaviour disorder” and reminds us that these behaviours are a challenge to service providers. self-injurious behaviour is considered

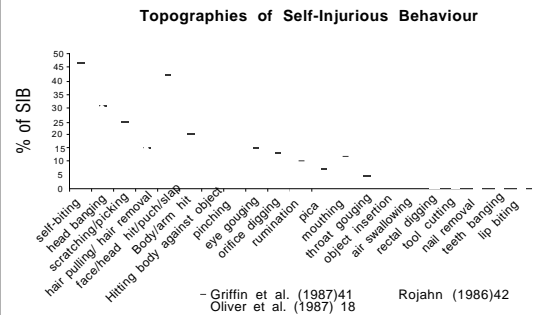
as one form of challenging behaviour along with other behaviours such as aggression and damaging the environment for example. Another term, stereotyped behaviour usually refers to meaningless, repetitive behaviours that have no immediate goal, such as rocking, hand-waving and spinning. Generally, these are different from self-injurious behaviour as they do not result in injury.

Finally, the term compulsive behaviour is used to describe behaviours that appear to be, to some extent, out of the control of the individual and the individual appears driven to show the behaviours. The reason for mentioning these two types of behaviour is that self-injurious behaviour is sometimes referred to as stereotyped or compulsive. This tends to mean that the self-injury is either repetitive, meaningless and not goal directed or that the self-injury appears to be out of the individual's control and the person appears driven to show the behaviour. (Compulsive self-injurious behaviour is discussed in Chapter 5). It should be noted that these terms are often used carelessly and it is advisable to be cautious about inferring anything about the cause of self-injurious behaviour from their use.

3.2: How common is self-injurious behaviour?

Within the population of people with intellectual disability the prevalence of self-injurious behaviour varies depending on how the study was conducted. Differences in the definitions that have been used, the time period within which the behaviour should occur to be counted, whether the information was collected by questionnaire or observation and the population that is considered will all influence the final estimate. When the largest studies are considered and studies are only included if they have a similar definition, time period and methodology and they include people with intellectual disabilities in a given geographical area (rather than just those in hospitals for example) then the prevalence of self-injurious behaviour is estimated to be 4-10%⁴. However, it is very clear that the prevalence of self-injurious behaviour is related

Box 3.2: Forms of self-injurious behaviour



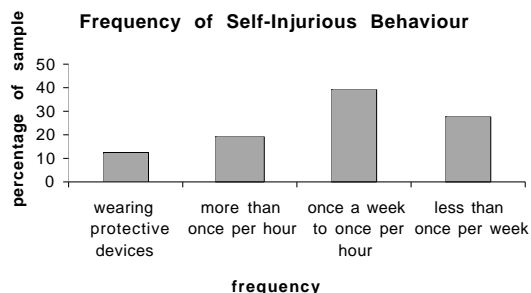
There are many topographies of self-injurious behaviour and one individual may show several different topographies. The graph above shows topographies of self-injurious behaviour seen in three prevalence studies of groups of people with intellectual disabilities.

to individual characteristics and we have begun to think of these as risk markers for self-injurious behaviour.

3.3: Forms of self-injurious behaviour

Self-injurious behaviour in people with intellectual disability can take a variety of forms. The most common are scratching or picking, biting and head hitting or banging (see Box 3.2) and there is enormous variability in the severity of self-injurious behaviour. Behaviours can be very mild in that the act does not incur immediate damage e.g. a soft face slap, or very severe such that the single act can cause significant injury e.g. a head bang to the sharp corner of the table. How frequently the behaviour occurs is also extremely variable, from a small number of incidents in a month to behaviours which can occur many times in an hour. (see Box 3.3) Additionally the temporal pattern of self-injurious behaviour can vary. Some self-injurious behaviour occurs in discreet bursts i.e. a bout of head-banging may be limited to 10 or 20 headbangs in a day, all occurring within the space of a minute or two. Alternatively, the behaviour may occur at a low level throughout a day, every few minutes.

Box 3.3: How frequent is self-injurious behaviour?



Oliver *et al.* (1987)¹⁸ carried out a total population survey of self-injurious behaviour in individuals with an intellectual disability in one health region of the UK. Six hundred and sixteen adults and children were found to have engaged in self-injurious behaviour sufficient to have caused tissue damage in the previous four months. 596 of these were screened and the frequency of self-injurious behaviour recorded in this group is shown in the graph above.

Another way in which the pattern may vary is that self-injurious behaviour may be problematic for weeks or months and then disappear or occur at a much lower level for a similar period. This pattern may be repeated over time. Whilst the importance of these different patterns is at pres-

ent unclear, it is possible to speculate on what these may tell us about the reasons for self-injurious behaviour and this is considered in more detail in Chapter 6.

3.4: Risk markers for self-injurious behaviour

The strongest risk marker for self-injurious behaviour is the degree of intellectual disability and this finding has been replicated across numerous studies. It is now very clear that the greater the degree of intellectual disability then the higher the prevalence of self-injurious behaviour (see Box 3.4). We have conducted a recent analysis of all prevalence studies and this indicates that the probability of showing self-injurious behaviour in children and adults who have severe or profound intellectual disability is more than four times higher than for those who have a moderate or mild intellectual disability⁵.

Another important risk marker is the presence of a genetic syndrome. Children and adults who have Lesch-Nyhan Syndrome almost always show self-injurious behaviour, primarily by biting their fingers and lips⁶. This one-to-one relationship between a genetic syndrome and self-injurious behaviour is not found in any other genetic syndrome. However, it is clear that the prevalence of self-injurious behaviour is much

Box 3.4: How is the prevalence of self-injurious behaviour influenced by intellectual disability?

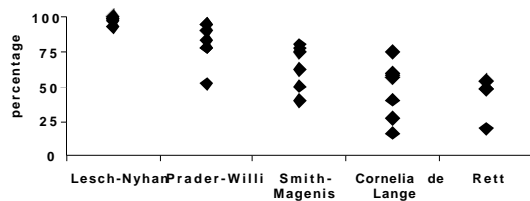
Several studies have examined the effect of degree of learning disability on the prevalence of self-injurious behaviour. The studies outlined in the table show that the prevalence of self injury increases as the degree of learning disability becomes more severe.

	Borderline (IQ $-68+$)	Mild (IQ 52-67)	Moderate (IQ 36-51)	Severe (IQ 20-35)	Profound (IQ 0-19)
Ballinger (1971) ⁴³	3.9	5.6	10.4	17.8	29.4
Ross (1972) ⁴⁴	—	13	18	25	26
Jacobson (1982) ⁴⁵	—	1.8 (younger) 2.9 (older)	3.4 (younger) 3.4 (older)	9.1 (younger) 6.4 (older)	14.3 (younger) 18.0 (older)
Maisto <i>et al</i> (1978) ⁴⁶	—	0	3.4	-----20.7-----	

higher than chance in Prader-Willi⁷, Cri du Chat⁸ and Smith-Magenis⁹ syndromes for example (see Box 3.5). For some of these syndromes it is clear that it may be the risk factor of severe or profound intellectual disability that contributes to the higher prevalence e.g. Cri du Chat Syndrome. However, for others this risk factor is not present and an alternative explanation needs to be sought e.g. Prader-Willi and Lesch-Nyhan Syndromes. The extent to which there is an association between self-injurious behaviour and Cornelia de Lange Syndrome has been the subject of some debate. Our recent research has focused on this question and this is discussed in more detail in Chapter 4.

There is some evidence that autism is associ-

Box 3.5: Prevalence of self-injurious behaviour in genetic syndromes



Self-injurious behaviour is common in several genetic syndromes, including Cornelia de Lange. Data from various studies are shown above. Each datum point represents the result of one study of a syndrome. As can be seen, the prevalence rate varies across studies. The prevalence rate of self injury in individuals with intellectual disability who do not have a syndrome is 4-10%. In all cases above the prevalence is much higher, up to eight times higher in Lesch-Nyhan syndrome and two to four times higher in Cornelia de Lange syndrome (see Chapter 4).

ated with self-injurious behaviour¹⁰. However, some of the criteria for diagnosing autism do include the presence of self-injurious behaviour. It is not clear therefore, whether the diagnosis of autism has been given because an individual shows self-injurious behaviour or because there

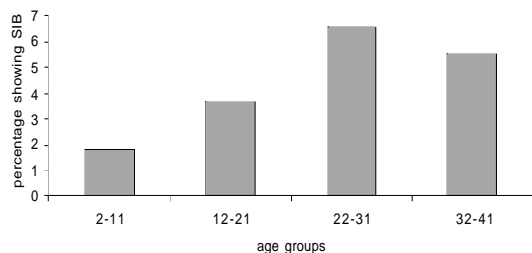
is genuinely an association between autism and self-injury. Autism is characterised by the so-called “Triad of Impairments” namely, impairments of socialisation, communication and imagination accompanied by the presence of repetitive and restricted behaviour¹¹ (see Box 2.7). The potential significance of an association between autism and self-injurious behaviour may lie with neurotransmitter disturbance that is assumed to occur in autism or the commonly associated intellectual disability. Additionally, it has been argued that individuals who have autism may show repetitive behaviours because there is an impairment of executive function¹² and thus limited control of the initiation of behaviours and the termination of behaviours (see Box 3.6). This is put forward as an explanation for the repetitive behaviours that are observed in autism such as stereotyped, compulsive and ritualistic behaviours and, for some people self-injurious behaviour. There may be another significance to the association between autism and self-injurious behaviour in that stereotyped behaviours, which are common in autism, may precede the development of self-injurious behaviour (see Chapter 5)¹³.

Box 3.6: What is executive function?

Executive functions are mental processes that help us to control our behaviour. There are several features of executive functions, for example: planning, holding a mental representation in short term memory, and being able to inhibit an inappropriate response. This latter feature has led some authors to link problems with executive function-executive dysfunction to attention-deficit-hyperactivity-disorder (ADHD) as individuals with ADHD find it difficult to inhibit ongoing responses and have difficulty maintaining attention. It has also been recently proposed that executive dysfunction may play a role in some of the deficits of functioning seen in children and adults with autism. This may in part explain the repetitive and inflexible behaviours shown by many individuals with autism.

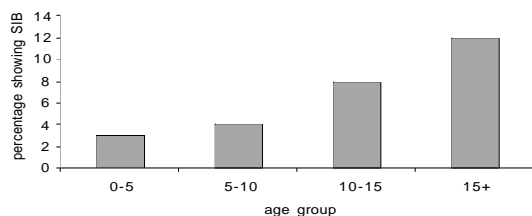
Box 3.7: The prevalence of self-injurious behaviour across the lifespan.

Figure 1: Kebbon & Windahl (1986)



Kebbon and Windahl (1986)⁴⁷ and Oliver *et al.* (1987)⁴⁸ both reported on the prevalence of self-injurious behaviours across various age groups. As can be seen above, self-injurious behaviour increases steadily with age until adolescence (Oliver *et al.*) and then falls off in adulthood (Kebbon and Windahl).

Figure 2: Oliver *et al.* (1987)



It has been argued that an expressive communication deficit is a risk marker for self-injurious behaviour¹⁴. More specifically, it is suggested that if expressive communication is significantly poorer than receptive communication then the risk is increased. However, it is extremely difficult to disentangle poor expressive communication from the degree of intellectual disability because many people with profound and severe intellect disability will have significant expressive communication problems. Consequently, much research in this area is confounded. However, it may well be that poor expressive communication is a critical contributor to the development of self-injurious behaviour and the reason for this is discussed in Chapter 5.

There is some limited evidence that both physical disability and sensory disability are risk

factors for self-injurious behaviour^{15, 16}. However, the studies that have reported this association have never really been replicated and, similar to the expressive communication risk factor, there is nearly always overlap with degree of intellectual disability. However, there does appear to be an association between vision impairment and self-injurious behaviour that is directed toward the eye¹⁷. This is further discussed in Chapter 5.

The final risk marker is age. From our past research, and similar research in Europe and the United States, it is clear that the prevalence of self-injurious behaviour rises with age up until the mid-twenties (see Box 3.7)¹⁸. It does appear that the development of self-injurious behaviour primarily occurs between the ages of approximately seven and mid-teens. However, this conclusion does depend on studies that have defined self-injurious behaviour by the resultant tissue damage. It is possible, and there is some research to support this, that the self-injurious behaviour does occur at a very young age¹⁹ but it is not recognised as self-injury because there is no tissue damage due to the children being too small to incur injury.

In addition to these individual characteristics there are some behaviours which appear to be associated with self-injurious behaviour. There is evidence that stereotyped, compulsive behaviours and movement disorders are more common when self-injury is present^{20, 21}. The significance of this is at present unclear. It maybe that stereotyped behaviour can evolve into self-injurious behaviour over time (see Chapter 5) or that the presence of these types of behaviours and movement disorders indicate a fundamental problem with control of movements²² (this is thought to be located in the basal ganglia see Box 3.8) and this is associated with self-injurious behaviour.

In combination these risk markers enable us to identify children who may be at higher risk of developing self-injurious behaviour. We would argue that children with a greater degree of intellectual disability, poor expressive communication and stereotyped or compulsive

behaviours are at higher risk for developing self-injurious behaviour. Additionally, some syndromes indicate a higher risk for the development of self-injurious behaviour and for these syndromes these other risk markers may be less relevant. At present there is no reason to think that these risk factors are not relevant to self-injurious behaviour in children with Cornelia de Lange Syndrome, consequently children who have these risk factors may be considered to be at greater risk for developing self-injurious behaviour.

3.5: Overview of the causes of self-injurious behaviour

The theories of the causes of self-injurious behaviour can be broken down into two broad areas. Biological theories emphasise fundamental neurological and medical factors as influential in the development of self-injurious behaviour. Conversely, psychological theories

Box 3.8: The basal ganglia and self-injurious behaviour

The term basal ganglia refers to a collection of structures of the brain including the striatum and globus pallidus. Historically, research around this area of the brain has been restricted to examining its role in movement disorders such as Parkinson's disease and Tourette's syndrome. In both Parkinson's disease and Tourette's Syndrome movement control is disordered. However, the high prevalence of self-injurious behaviour in individuals with Tourette's syndrome and the association of self-injurious behaviour with other repetitive movements or movement disorders suggests that there may be a common brain pathway involving the basal ganglia. Damage to the basal ganglia during development may therefore lead to motor stereotypies, self-injurious behaviour or movement disorder. This theory is supported by the increased levels of self-injurious behaviour in individuals with intellectual disability and specific genetic disorders that affect early brain development.

consider the environment to be important, particularly with regard to the development of self-injurious behaviour. Whilst these theories tend to be clearly separated in the research literature there is no evidence that they are mutually exclusive and, in genetic syndromes such as Cornelia de Lange, interactions between aspects of the theories might help towards a better understanding of self-injurious behaviour^{23, 24}.

3.5.1: Neurotransmitter or neuromodulator dysfunction

Neurotransmitters provide a chemical link between the ends of the individual nerves that make up our brain and the rest of our nervous system. Different neurotransmitters tend to be associated with different functions and three neurotransmitters have been implicated as relevant to self-injurious behaviour on the basis of animal research, pharmacological intervention and, more recently, MRI studies (see Box 3.9)

Dopamine. Dopamine is clearly implicated in movement and the effect of dysfunction of dopamine is most clearly seen in Parkinson's disease. There is some evidence that dopaminergic dysfunction is related to self-injurious behaviour and stereotyped behaviours²⁵. However, at present medication that targets dopaminergic dysfunction has not been reliably demonstrated to influence self-injurious behaviour²⁶.

Serotonin. Serotonin dysfunction is implicated in both depression and compulsive behaviours and there is some evidence that the use of serotonergic agents, and more specifically SSRI's (selective serotonin reuptake inhibitors) influence self-injurious behaviour, interestingly by both decreasing and increasing the behaviour. Similar to the dopamine story at present there is no reliable evidence that the use of serotonergic agents can influence self-injurious behaviour.

Endorphins. Endorphins are the body's natural

Box 3.9: Biological research into self-injurious behaviour

Animal studies: For many years scientists have tried to develop an animal model of self-injurious behaviour in an attempt to both understand the behaviour and evaluate treatments for it. It has been found that injecting rats with certain chemicals, such as amphetamines, can induce self-injurious behaviour.

Pharmacological intervention: If the theory that (at least in some individuals) self-injurious behaviour is caused by neurotransmitter disturbance is true, then a drug treatment would be the most appropriate solution. Several studies have been carried out using various different drugs to treat self-injurious behaviour but results have been inconclusive as yet.

Magnetic Resonance Imaging studies: MRI scanners give high resolution images of the brain (a bit like an x-ray). Scientists have recently used these techniques to look at the brains of individuals with Lesch-Nyhan syndrome to see if there are any anatomical or functional differences that might be an indication of what causes the behaviour. It has been found recently that certain parts of the brain in individuals with Lesch-Nyhan syndrome (nearly all who have this syndrome show self-injurious behaviour) were significantly smaller than in individuals who do not have Lesch-Nyhan syndrome.

painkillers and are similar in chemical structure to morphine and are released at times of great stress and when pain is experienced (see Box 3.10). It has been suggested that in people who shows self-injurious behaviour endorphin production might be disturbed such that two things happen. First, it has been suggested that individuals are releasing too much endorphin and consequently they are not experiencing pain when they self-injure²⁷. Second, it has been suggested that people may become addicted to their own endorphins and consequently self-injure in order to both gain a euphoric effect (similar to that caused by opiate use) and avoid the negative consequences of withdrawal from their own endorphins. The evidence for both of these theories is rather weak and tends to rely on the observation that giving medication that effectively blocks endorphins can lead to a reduction in self-injurious behaviour in some people (see Box 3.11). However, it can be argued that this medication works by simply increasing the pain that is experienced when self-injury occurs and consequently the behaviour decreases (this is related to the idea of response efficiency, see Chapter 7). This explanation does not necessarily require there to be existing dysfunction in the production of endorphins.

Box 3.10: What are endorphins?

Endorphins are neuromodulators found in the brain that have pain-relieving properties similar to morphine. Endorphins interact with opiate receptor neurons to reduce the intensity of pain. Among individuals afflicted with chronic pain disorders, endorphins are often found in high quantity. The effect of endorphins appears to be responsible for the so called “runner’s high”, the temporary loss of pain when severe injury occurs, and the analgesic effects that acupuncture and chiropractic adjustments of the spine offer. Besides behaving as a pain regulator, endorphins are also thought to be connected to physiological processes including euphoric feelings, appetite modulation, and the release of sex hormones.

3.5.2: Medical conditions

There is some evidence in the literature that medical conditions such otitis media²⁸ (middle ear infections) skin infections²⁹ and irritation can lead to self-injurious behaviour in children and adults who have intellectual disability. (see Box 3.12) More recently it has been suggested that

people who experience gastro-intestinal reflux also show self-injurious behaviour³⁰. The relevance of this research to Cornelia de Lange Syndrome is clear given the health problems that we described in Chapter 2 and is discussed further in Chapters 5, 6 and 7.

3.5.3: Psychological factors

Psychological theories of the cause of self-injurious behaviour in people with intellectual disabili-

ty have dominated the research literature and are supported by a vast array of empirical research studies^{31, 24}. The dominant perspective within the psychological theories is that of operant learning theory which proposes that self-injurious behaviour is a learned behaviour that occurs because it is rewarded (or reinforced) by either sensory or social consequences³². The evidence for this perspective is very strong and has been built up over a period of forty years. However, the primary issue is the extent to which this perspective is

Box 3.11: Case studies of drug trials in self-injurious behaviour

Naltrexone and naloxone are opioid antagonists that work by blocking the uptake of endogenous opioids or endorphins. It has been suggested that rather than environmental reinforcers such as attention, self-injurious behaviour in some individuals is reinforced by the “high” produced when the endogenous opioids are released after self-injurious behaviour has occurred. Opioid antagonists such as naloxone and naltrexone block this release of endorphins and the individual therefore does not experience the “high”. Many studies have attempted to demonstrate the efficacy of these drugs in reducing self-injurious behaviour but, as highlighted by the studies below, the results have been inconsistent.

Study	Drug tested	Results
Sandman et al (1983) ⁴⁸	Naloxone	Substantial suppression of self-injurious behaviour but only whilst drug active in system.
Beckwith et al (1986) ⁴⁹	Naloxone- various doses	No effect on rate of self-injurious behaviour, regardless of dose given.
Luiselli et al (1989) ⁵⁰	Naltrexone	No effect on multiple forms of self-injurious behaviour.
Symons and Thompson (1998) ⁵¹	Naltrexone both alone and with FCT*	Naltrexone alone produced a 50% reduction in self-injurious behaviour.

*FCT= Functional communication training (see Chapter 7)

One study of naloxone treatment (Richardson and Zaleski, 1983⁵²) reported an increase in SIB during drug administration which decreased again when medication was discontinued. They explained this increase in terms of an extinction burst. Extinction occurs when a normal reinforcer of a behaviour (i.e.attention) is removed (i.e. you ignore the behaviour instead). It is common for an extinction burst (a subsequent, often dramatic increase in the target behaviour) to occur as the individual strives to find reinforcement as usual (se Box 7.13). The subsequent decrease in self-injurious behaviour when drug administration was stopped might therefore be explained by the “re-activation” of the reinforcer.

applicable to all people who show self-injurious behaviour at all times. Certainly, there is very good evidence that when assessment procedures show that a behaviour is learned and is occurring because of reinforcement then interventions can be very effective³³. Given this very strong evidence base we will focus on this psychological approach in this book.

For the purposes of understanding self-injurious behaviour and the assessment process it is useful to break the operant learning theory into two components and consider the role of sensory reinforcement and social reinforcement separately.

3.5.3.1 Sensory reinforcement

The basis to the sensory reinforcement argument is that self-injurious behaviour occurs because the immediate sensory or perceptual consequences of the behaviour are experienced as pleasant either by the presentation of positive sensation or the removal of an unpleasant sensation³⁴. The evidence for this part of the operant theory comes from two sources. First, when alternative forms of stimulation are provided³⁵, and particularly when the form of stimulation is similar to that associated with the self-injurious behaviour, then the behaviour tends to decrease, at least temporarily. Second, when the stimulation from the self-injurious behaviour is blocked, this is called sensory extinction³⁶, and then the self-injury tends to cease. This theory has also been employed to understand stereotyped behaviours.

3.5.3.2 Social reinforcement

Since the late 1960s the evidence has grown that self-injurious behaviour can occur because it leads to social reinforcement (reward) from people who interact with the person showing the behaviour. In the last decade there has been a greater understanding of how this process unfolds and why it occurs²⁴. Broadly there are two ways in which the process operates at any one point in time. First, self-injurious behaviour

Box 3.12: Self-injurious behaviour associated with medical conditions

Self-injurious behaviours such as head banging are often reported in normally developing children (15-20%). De Lissovoy²⁸ examined this behaviour in a series of papers in the early 1960's and found that the children in his study that were head banging were significantly more likely to have had otitis media (middle ear infection) than children in a matched comparison group. A more recent example of this association comes from Colville and Mok (2003)³³ who describe two children with no intellectual disabilities who began to show self-injurious behaviour whilst in hospital. Both children were on ventilators in an intensive care ward and both began to show lip-biting behaviour. In both cases, the behaviour was extinguished through increased psychosocial input and anxiety reduction.

can occur because it leads to the presentation of positive social contact or more tangible things such as food, drink or the presentation of activities³⁷. Second, self-injurious behaviour can occur because it leads to an unpleasant or aversive social contact being removed³⁸. The form of this unpleasant social contact is usually the presentation of tasks or demands that the individual simply does not want to do or finds too difficult or unrewarding. An understanding of this process of social reinforcement is critical to effective intervention consequently we have allocated much of the space on psychological assessment and intervention to this area.

It is important to note that the theories that have been developed that are described do not necessarily apply to all individuals at all times. Consequently, we emphasise the importance of assessment procedures to uncover which causes might be applicable to a given individual at any one point in time in order to guide the intervention process. However, it is equally important to recognise that for any individual more than one cause might be influential and it is almost certainly the case that causes may change over time.

This is particularly true for socially reinforced self-injurious behaviour. For this theory to be correct the behaviour has to be occurring in the first place in order for it to be reinforced. It is most likely therefore that self-injurious behaviour initially occurs in response to a minor illness or as a form of stereotyped behaviour that is reinforced by the sensory stimulation and then comes to be socially reinforced by those who interact with the person showing the behaviour^{39,13}. Our past research has shown us some evidence that this is the case in children with severe intellectual disability and there is no reason to think that this is not applicable to children with Cornelia de Lange Syndrome.

3.6: Summary

Self-injurious behaviour is associated with a greater degree of intellectual disability and some specific genetic syndromes. The prevalence of self-injury increases with age until the mid-20's and the most common forms of self-injury are biting, picking and striking. Theories of the causes of self-injury in all people with intellectual disability are of two main types: biological and psychological. Biological theories tend to focus on possible neurotransmitter dysfunction and the role that might be played by minor illnesses, pain and discomfort. Psychological theories are concerned with the sensory stimulation that follows self-injury and the possible rewarding responses by carers. These theories seem to be applicable to children and adults with Cornelia de Lange Syndrome who show self-injurious behaviour and are explored in detail in subsequent chapters.

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Chapter 4: Self-injurious behaviour in Cornelia de Lange Syndrome

4.1: Introduction

Since Dr. William Nyhan's early reports that self-injurious behaviour might be associated with Cornelia de Lange Syndrome (Nyhan, 19721), the syndrome has generally been known as one in which self-injury frequently occurs and it has often been argued that the behaviour may have a biological cause. As a result of these assertions, two types of studies have attempted to see if an association does indeed exist between Cornelia de Lange Syndrome and self-injurious behaviour.

First, there have been studies that have reported on relatively small numbers of people with Cornelia de Lange Syndrome. Many of these studies have looked at patients in clinical settings (see Box 4.1). Also within this category of 'small scale studies', are the many single case reports that have been published in academic journals. These have simply described children and adults with Cornelia de Lange Syndrome and the self-injurious behaviour that they display. There are two problems with this type of small scale research. As many of these studies include only individuals who were being seen in clinical settings, it is possible that they may have been seen or referred to the authors precisely because they showed self-injurious behaviour. This would have the effect of elevating the prevalence figures of self-injury.

Secondly, with regard to the individual case reports, because of both Nyhan's early reports that Cornelia de Lange Syndrome and self-injury are associated, and the intriguing nature of self-injury which has caused a great deal of academic interest, it is possible that a bias has emerged. Due to this, researchers may have been more

likely to observe and describe individuals with Cornelia de Lange Syndrome who engage in self-injury, publishing reports that in turn make the association look stronger. Consequently, the combination of Nyhan's early reports associating Cornelia de Lange Syndrome and self-injurious behaviour, together with the studies in clinical settings, and the individual case studies, has tended to give the impression that self-injurious behaviour was shown by relatively high numbers of people with Cornelia de Lange Syndrome. Given the limitations of this research, it is important to also look at the results from the second type of study, large scale surveys.

Following on from the small-scale studies, several large surveys have been conducted, usually via the Cornelia de Lange Syndrome Foundation. Such studies are less likely to be biased, because they aim to recruit as many different people with Cornelia de Lange Syndrome as possible, regardless of the behaviours they display. As a result, this type of research is perhaps the best indicator of the true prevalence of self-injurious behaviour in Cornelia de Lange Syndrome. However, the prevalence of self-injury in these large-scale surveys is, in fact, similar to that seen in the smaller studies, approximately 60%. This figure of 60% shows us very clearly that there is not a one-to-one relationship between the syndrome and self-injurious behaviour (see Box 4.2).

4.2: Comparing self-injury in Cornelia de Lange Syndrome with self-injury in intellectual disability

Even though the prevalence of self-injurious behaviour in Cornelia de Lange Syndrome is cer-

Box 4.1: Studies reporting self-injurious behaviour in individuals with Cornelia de Lange Syndrome seen in clinical settings

Authors	Summary of the study	Prevalence of self-injury
Bryson, <i>et al.</i>(1971).²	The authors surveyed and observed all patients in a large state hospital with Cornelia de Lange syndrome (7) and then observed those in whom self-mutilation was a major feature.	57.1% of Cornelia de Lange syndrome patients in the hospital displayed 'self-mutilation'.
Greenberg & Coleman (1973).³	The study investigated serotonin levels in 11 patients with Cornelia de Lange syndrome. However, behavioural symptoms of the participants were also noted.	27.2% were noted to show self-injury.
Johnson <i>et al.</i> (1976).⁴	The authors looked closely at the 'behavioural phenotype' of 7 patients with Cornelia de Lange syndrome using videotape observations.	57.1% displayed at least form of self-injurious behaviour.
Beck (1987)⁵	A psychosocial assessment of 36 patients with Cornelia de Lange syndrome was conducted. Information was gathered from informants and behavioural difficulties were recorded.	16.7% showed 'behaviour problems in the form of self-mutilation'

tainly higher than that in all people with intellectual disability (5-10%, see Chapter 3) this still does not necessarily mean that self-injurious behaviour is any more common in Cornelia de Lange Syndrome than we would expect by chance. The reason for this is that some of the risk markers for self-injurious behaviour that are seen in all people with intellectual disability (see Section 3.4) might also be evident or more common in those individuals with Cornelia de Lange Syndrome who show self-injurious behaviour. It may be these risk markers, rather than the syndrome itself, that leads to a high prevalence of self-injurious behaviour.

In order to determine whether self-injurious behaviour is more common in Cornelia de Lange Syndrome than we would expect by chance, we conducted a study in which we compared 54 people with Cornelia de Lange Syndrome with 46 people without the syndrome. We aimed to recruit as many people with Cornelia de Lange Syndrome into the study as possible and contact-

ed families through a number of sources. Following this, we recruited individuals without Cornelia de Lange Syndrome and matched the groups in terms of age, gender, level of mobility and degree of intellectual disability. By doing this, we were trying to ensure that any risk markers for self-injury were the same for both groups. We visited participants in their day-care settings, and obtained further information through both questionnaires and interviews with parents, carers and teachers. We asked whether self-injurious behaviour had occurred in the previous month. Self-injurious behaviour was defined as 'non accidental behaviours producing temporary marks or reddening of the skin, or cause bruising, bleeding or other temporary or permanent tissue damage'.

When we did this, we found that 55.6% of those with Cornelia de Lange Syndrome had showed self-injurious behaviour, whilst 41.3% of those in the comparison group had displayed the behaviour. This showed us that in people

Box 4.2: Survey studies of self-injurious behaviour in Cornelia de Lange Syndrome

Authors	Summary of the study	Prevalence of self-injury
Hawley <i>et al.</i> (1985).⁶	64 families caring for people with Cornelia de Lange syndrome completed a questionnaire. The study investigated the natural course of and problems in Cornelia de Lange syndrome, as well as its mode of inheritance.	57 % ‘manifested behaviour management problems, including self-injurious behaviour.’
Gualtieri (1990).⁷	131 carers of people with Cornelia de Lange syndrome completed a questionnaire investigating behaviour in Cornelia de Lange syndrome .	‘88 respondents had current problems with self-injurious behaviour, 10 more had past, possible or rare self-injurious behaviour, and 33 were reported as having had none.’ 74.8%
Sarimski (1997).⁸	Communication, social-emotional development and parenting stress were assessed in 27 families caring for people with Cornelia de Lange syndrome through a postal survey.	40.7% of subjects displayed self-injurious behaviours’
Berney <i>et al.</i> (1999).⁹	A postal questionnaire was distributed to 49 carers in order to investigate the behavioural phenotype in Cornelia de Lange syndrome .	56% showed ‘self-injury occurring at least occasionally’
Hyman <i>et al.</i> (2002).¹⁰	A postal questionnaire was completed by 86 carers in order to look at the association between self-restraint, self-injurious behaviour and compulsions in Cornelia de Lange syndrome.	62.8% displayed self-injurious behaviours in the past month.

with Cornelia de Lange Syndrome, self-injurious behaviour was more likely to occur than for people of the same age, gender and degree of disability who did not have the syndrome. However, this still does not necessarily mean that the behaviour is associated with Cornelia de Lange Syndrome. It could be that this 14.3% difference is due to chance (see Box 4.3). In fact, when we conducted statistical tests to examine the significance of this finding we found that there was no significant difference between the two groups. This means that when we control for the risk markers, self-injurious behaviour to the point of tissue damage, is no more likely to occur in Cornelia de Lange Syndrome than in

other people who have the same degree of intellectual disability, age and gender. Further analyses also showed that having a diagnosis of Cornelia de Lange Syndrome does not predict the presence of self-injury above the risk markers and characteristics that are associated with self-injury. In other words, if two individuals are of similar age, gender, level of mobility and degree of learning disability and have other similar characteristics otherwise associated with self-injury, but one has Cornelia de Lange Syndrome and one does not, there would be no reason to predict that the person with Cornelia de Lange Syndrome would be more likely to engage in self-injurious behaviour.

Box 4.3: Chance levels in statistics

In scientific research, statistical tests are carried out to determine whether chance can reasonably explain the differences found between two groups. In other words, a calculation determines the probability that the results observed could be obtained if there was no real difference between the groups. Groups are considered significantly different from one another if the probability of obtaining the difference by chance alone is no greater than 5% (0.05). However, when the probability is 5% or greater, chance cannot be ruled out as an explanation for the difference.

This finding is perhaps surprising given the past research and the reported association between the syndrome and self-injury. However, the study does differ from previous research in one important way. This is that we compared the prevalence of self-injurious behaviour in Cornelia de Lange Syndrome with a similar group people with intellectual disability, but without the syndrome. It is only when we do this that we can see that the self-injury in Cornelia de Lange Syndrome is associated more to the intellectual disability and other risk markers for the behaviour than the syndrome itself. However, although we would now argue that self-injurious behaviour to the point of tissue damage is not more common in Cornelia de Lange Syndrome than in people with a similar degree of intellectual disability it is still a concern that approximately 60% of people with Cornelia de Lange Syndrome do show self-injurious behaviour.

4.3: Forms and site of self-injury in Cornelia de Lange Syndrome

In order to further understand the self-injurious behaviour that we see in Cornelia de Lange Syndrome, we visited and observed individuals with and without the syndrome over the course of a day in their usual day-care environment.

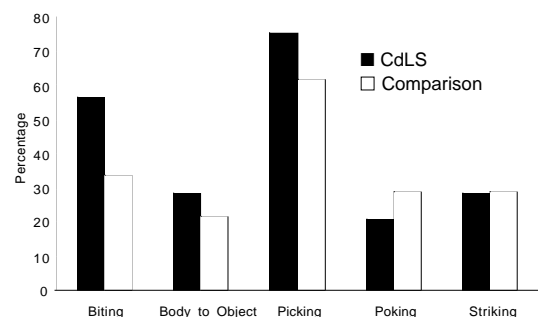
Following this, we were able to examine the severity of self-injurious behaviour in the two groups and look at the types of self-injurious behaviour they displayed, together with the part of the body to which the behaviour was directed.

The comparisons conducted on the severity of self-injury and the frequency of self-injury showed no differences between the two groups. We also found no difference between the types of self-injurious behaviour shown by people with and without Cornelia de Lange Syndrome, except for the biting which was significantly higher in people with Cornelia de Lange Syndrome (see Box 4.4). Additionally, there were no differences between the two groups in terms of the part of the body to which self-injurious behaviour was directed, except that people with Cornelia the Lange Syndrome were more likely to injure their hands than the comparison group (see Box 4.5).

So, whilst the prevalence of self-injury is not

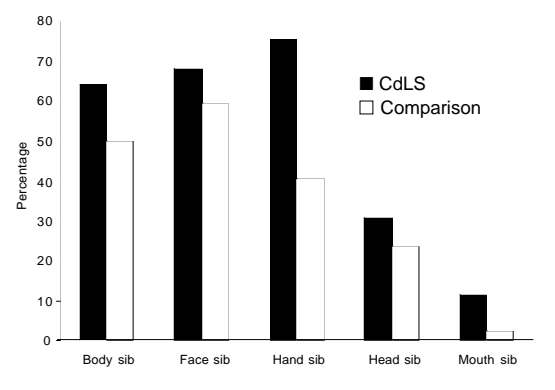
Box 4.4: Types of self-injurious behaviour in Cornelia de Lange syndrome

The graph below indicates the frequency of different types of self-injury that were observed in individuals with Cornelia de Lange syndrome and individuals with intellectual disability without the syndrome. The only significant difference between these two groups with regard to the type of self-injury displayed was that individuals with Cornelia de Lange syndrome were more likely to engage in biting than the comparison group.



Box 4.5: Site of self-injurious behaviour in Cornelia de Lange Syndrome

The graph below indicates the parts of the body that were the most frequent sites of self-injurious behaviour in individuals with Cornelia de Lange Syndrome compared to individuals with intellectual disability without the syndrome. The only significant difference between these two groups with regard to site of injury was that individuals with Cornelia de Lange Syndrome were more likely to injure their hands than the comparison group.



significantly higher in Cornelia de Lange Syndrome than a comparison group, it is clear that children and adults who have Cornelia de Lange Syndrome are more likely to self-injure by biting and the site of injury is more likely to be on their hands. The significance of this is unclear but this may be related to peripheral sensory neuropathy (see Section 2.3) and this is further discussed in Chapter 5.

4.4: Self-injury, compulsive behaviours and self-restraint in Cornelia de Lange Syndrome

In a further analysis of studies we have conducted we have been able to show that in Cornelia de Lange Syndrome there is an association between self-injurious behaviour, the presence of compulsive behaviours and self-restraint (see Box 4.6). This finding is of interest because it may indicate that for some people with Cornelia de Lange Syndrome, the self-

injurious behaviour has a compulsive quality (see Chapter 2) and thus the behaviour is to some extent out of the individual's control. Under these circumstances the person may show self-restraint or develop a preference for protective devices in order to assist with the control of their behaviour. In a second study that we conducted we did find that some behaviours were associated with self-injurious behaviour in both Cornelia de Lange Syndrome and our comparison group. These behaviours were: stereotyped behaviour, compulsive behaviour and hyperactivity. The presence of these three types of behaviours predicted the presence of self-injurious behaviour. This is of interest because it may indicate that there is a motor disorder or a problem with behavioural inhibition that underpins all of these behaviours in Cornelia de Lange Syndrome. Recently it has been speculated that the area of the brain that might be responsible for this association and these motor disorders is the Basal Ganglion (see Section 3.4 and Box 3.6).

Box 4.6: The association between self-injurious behaviour, compulsive behaviour and self-restraint in Cornelia de Lange Syndrome

A study by Hyman *et al.* (2002) considered the relationship between self-injurious behaviour, compulsive behaviour and self-restraint in individuals with Cornelia de Lange Syndrome. The study showed that 64.3% of individuals displaying self-injurious behaviour also engaged in self-restraint. In addition to this, significantly more compulsive behaviours were shown by individuals displaying self-injurious behaviour and self-restraint compared to those without these behaviours. The findings suggest that there is some sort of relationship between self-injury, self-restraint and compulsive behaviours. The precise nature and significance of this association is yet to be established.

4.5: Summary

Self-injurious behaviour is shown by approximately 60% of individuals who have Cornelia de Lange Syndrome. Interestingly, although this is a high figure, people with Cornelia de Lange Syndrome are not necessarily more likely to show self-injurious behaviour than people with the same degree of intellectual disability and other risk marker characteristics. However, the self-injury does differ in three potentially important ways. First self-injury is more likely to be directed towards the hands. Second, self-injurious behaviour is more likely to take the form of biting and third there is some tangential evidence that for some people with Cornelia de Lange Syndrome the behaviour appears to need active control by self-restraint.

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Chapter 5: Causes of self-injurious behaviour in Cornelia de Lange Syndrome

5.1: Introduction

When thinking about self-injurious behaviour the word “cause” is itself problematic. There is almost never a one-to-one relationship between what we think of as a cause of self-injurious behaviour and the behaviour itself. It is better to think of factors that make the behaviour more or less likely to occur. It is also important when thinking about self-injurious behaviour in children and adults who have Cornelia de Lange Syndrome that a variety of factors are considered and that it is not assumed that the cause of self-injurious behaviour at one time is necessarily the cause at a later date. There is very clear evidence that the causes of self-injurious behaviour differ between people and that they may change over time. Additionally, even if the form of self-injurious behaviour is similar for two people (e.g. they both bang their heads) it does not necessarily mean that it happens for the same reason. In this chapter we will consider the possible causes of self-injurious behaviour in Cornelia de Lange Syndrome, focussing first on internal causes and then moving on to external causes and then we will consider how these might interact at one point in time and over time.

5.2: Internal causes

It is useful to think about the causes of self-injury as being internal to the person, usually meaning things that we cannot see and external, things in the environment that seem to affect the behaviour. There are two types of internal causes, pain and discomfort and sensory stimulation, and we will describe these in turn.

5.2.1: Relief of pain or discomfort

It was noted in Chapter 3 that in people with intellectual disability, as well as those who do not have a disability, self-injurious behaviour can occur in response to painful medical conditions and discomfort. It was also noted in Chapter 2 that there are number of medical conditions in Cornelia de Lange Syndrome that can give rise to pain and discomfort. Whilst there is very limited research data on the relationship between pain and discomfort in Cornelia de Lange Syndrome and self-injurious behaviour, our clinical experience leads us to believe that this should always be considered first.

It is possible that the dental problems experienced by children adults with Cornelia de Lange Syndrome, such as decay associated with thin

Box 5.1: The experience of pain

There is very little attention paid to the experience of pain in people who have intellectual disability and show self-injury. This is curious because it is probably the first thing that would be investigated in someone who showed self-injury but did not have an intellectual disability or psychological disorder. It is clear that the experience of pain cannot be accounted for simply by suggesting that a nerve fires when there is a painful stimuli and the brain registers pain. There are other processes involved. Ronald Melzack and Patrick Wall, when describing their theory of pain, draw attention to how pain can be blocked by endorphins (the bodies natural opiates) or by physical stimulation (rubbing and scratching) that causes pain blocking nerves to fire.

enamel, reflux and the crowding of teeth, will lead to discomfort. This may in turn lead to self-injurious behaviour as the individual attempts to relieve the pain and discomfort by banging the teeth or area around the mouth. Certainly we know that in anyone physical stimulation of a local site of pain does relieve discomfort, at least temporarily (see Box 5.1)¹. The similar argument may be made for a tooth eruption and it should not be surprising if children and bang or bump the area around the mouth around this time.

Similarly middle ear infection and blocked sinuses or sinus infections can cause pain and discomfort and may lead to banging or rubbing of the area around ears and the upper cheek and bridge of the nose. There is little doubt that children and adults who suffer from reflux as a result of gastro-intestinal problems experienced significant discomfort when reflux occurs. Again the extreme discomfort and burning sensation in the chest and throat may lead to scratching, punching or hitting of these areas and children and adults may push their fingers or hands into the throat in an attempt to relieve the pain and discomfort. Indications that reflux is occurring and painful include these behaviours as well as excessive drinking, food avoidance, approach-avoidance behaviours prior to food (repeatedly sitting down to meals but then moving away), arching of the back (see Box 5.2) and various forms of difficult behaviour in the period following meals.

The eye problems that children adults with Cornelia de Lange Syndrome experience may also evoke temporary or longstanding self-injurious behaviour. If the tear-ducts are not functioning correctly, and consequently no tears are being produced, then it is probable that the surface of the eye will become dry and irritated and this may lead to the child or adult rubbing the eye in an attempt to relieve this unpleasant sensation. Additionally, it is also important to be aware that constant rubbing around the eyes may lead to the eyelashes rubbing on the surface of the eye and causing some discomfort. This is particularly important as children and adults with

Box 5.2: Back arching in Cornelia de Lange Syndrome

In 1976 Johnson and his colleagues reported that individuals with Cornelia de Lange Syndrome are ‘non-social either with family members or strangers’²⁴. They reported that children with Cornelia de Lange Syndrome had an unusual but characteristic response to being held by other people. Individuals were described as arching their backs or bodies away from the person holding them. This was interpreted to be rejection of physical contact by the individual, which led the authors to think that individuals with Cornelia de Lange Syndrome do not like any form of social contact. We now think that that this characteristic back arching is more likely to be a way of relieving the pain and discomfort related to gastro-intestinal reflux that are a feature of the syndrome rather than a way of escaping social contact.

Cornelia de Lange Syndrome and tend to have long and thick eyelashes.

The role of a potential peripheral sensory neuropathy is worth considering in some detail. The sensation of pain is dampened by a peripheral sensory neuropathy². Additionally, it is possible that peripheral sensory neuropathy gives rise to a tingling or mild burning sensation in the arms, hands and fingers³. If this is the case then individuals may respond to this sensation by scratching, picking or biting the area in which the tingling is occurring. When this happens in the absence of any pain the behaviour may become more severe than would otherwise be the case. It should be noted that there is only limited evidence to date that a peripheral sensory neuropathy is evident in people with Cornelia de Lange Syndrome⁴. However, this possibility cannot yet be ruled out.

5.2.2: Sensory reinforcement

There is some evidence that self-injurious behaviour does occur because it leads to a pleasant

Box 5.3: Rods and cones in the retina of the eye

Rods and cones are photosensitive cells in the retina that convert light energy into electrical nerve impulses. The rods and cones lead into the optic nerve which enables information to reach the brain for interpretation. Whilst rods and cones are usually sensitive to light, they will also fire off nerve impulses if they are physically stimulated by pressing the eye, which leads to increased pressure in the eyeball and, in turn, the rods and cones.

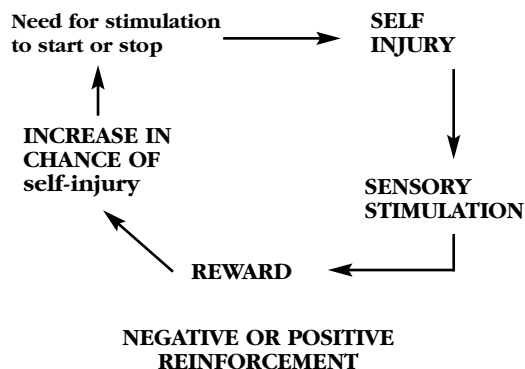
sensation that a person finds rewarding and consequently they will show the behaviour in the future ⁵. The clearest example of this is eye pressing and this commonly occurs when there is damage to the eye which causes the rods and cones in the rear of the eye to be unresponsive to light (see Box 5.3). Under these conditions physically pressing the eye will cause the rods and cones to fire and the individual will “see” flashes and patterns of light. It is not surprising therefore that when someone discovers this way of producing a sensation that they will reproduce it over long periods of time, especially when the eye is not producing any other form of stimulation from light.

There are of course a number of other ways in which the sensation that arises from mild forms of self-injurious behaviour might be rewarding. This effect might be enhanced when there is limited pain associated with the behaviour. Mild face slapping can produce a tingling sensation and pressing and banging the ears can result in “pleasant” noises.

It is also useful to think about the relief of pain and discomfort as a sensory reinforcement process. In this case the reward is not the presentation of something positive after the behaviour but the removal of an aversive or unpleasant sensation. The best example of this is scratching an itch. In this example the itch is scratched because the behaviour of scratching is rewarded by the itch subsiding.

The process of sensory reinforcement is shown in Box 5.4.

Box 5.4: Sensory reinforcement of self-injurious behaviour



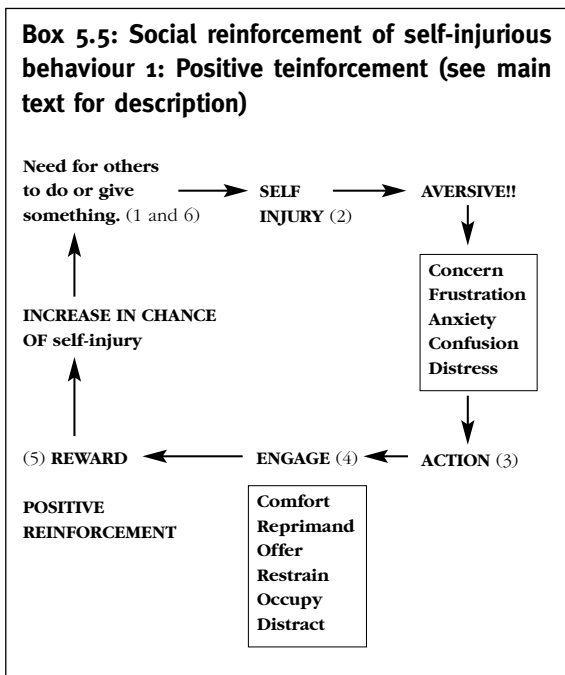
Some self-injurious behaviour may be reinforced by sensory reinforcement in which the behaviour itself provides the individual with reinforcement rather than another person in their environment. Take the example of a child with ear ache. The child has a pain and they therefore have a need for stimulation (i.e. the pain) to stop. In this example the self-injurious behaviour may be head banging. The head banging provides stimulation but also, more importantly, removes the pain (albeit temporarily). This is an example of negative reinforcement as the child is rewarded by the pain stopping and there is now an increased chance that, given an ear ache in future, the child will follow this same process again.

5.3 External causes

When we think about the external causes we mean factors that are in the environment that seem to effect the level of self-injury. The most important of these is the effect that the responses of others has on the self-injury and this usually takes two forms: presenting rewarding attention and removing unpleasant demands. We will describe these in turn.

5.3.1: Positive social reinforcement

As was discussed in Chapter 3 there is a good deal of evidence that self-injurious behaviour can be rewarded or reinforced by the presentation of social contact (attention) and it is important to understand how this process occurs^{6, 7, 8}. The process is shown in Box 5.5 and the numbers in the figure in Box 5.5. refer to the sequence of events that are listed here:



1. The child is on their own and has no stimulation. Initially, the child may show self-injurious behaviour either because it gives rise to pleasant sensory stimulation or because it relieves discomfort (see section 5.3.1) or as the end result of a stereotyped behaviour or it is simply a chance act (an accidental bump of the head).
2. The self-injury occurs and seen by another person (parent or carer).
3. The other person finds the self-injury unpleasant or aversive and consequently acts to stop the self-injury from recurring or tries to find out the reason for the self-injury.
4. The other person engages with the child who

has just shown self-injurious behaviour and whilst preventing further instances of self-injury and trying to find the cause, may comfort, distract or restrain the person or use any combination of these strategies.

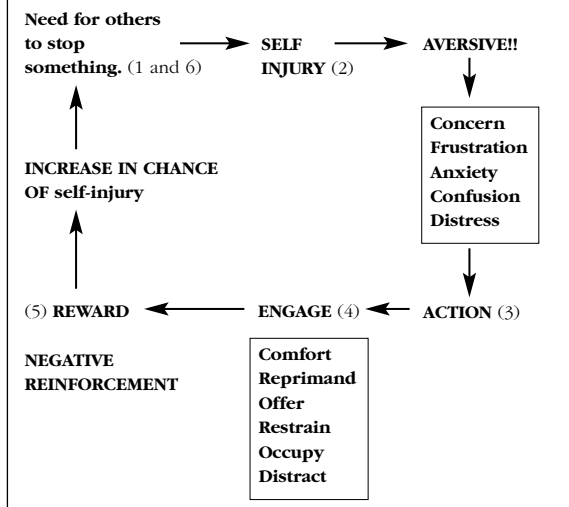
5. The child finds this contact with the other person pleasant and rewarding (reinforcing). This makes it more likely that the next time the person is alone and without contact they will self-injure.
6. The child is alone and has no stimulation (and thus motivated to seek contact). Self-injury occurs because in the past it has led to rewarding social contact with another person. (Go to 2 above).

Once this process has occurred an number of times the child will very quickly learn to self-injure because it leads to rewarding attention from another person. This is not to say that the child necessarily intends to injure him or herself or intends to gain the attention of someone else. It is an entirely natural process whereby a self-injurious behaviour is so unpleasant that it evokes an entirely natural reaction from another person and the contact with that person is also naturally rewarding or reinforcing.

5.3.2: Escape from task demands

The idea that self-injurious behaviour can occur because it leads to attention from other people is not a new one and was first put forward in the late 1960s⁹. It was some time later that it was also suggested that self-injurious behaviour could have a different kind of effect on other people which is that of reducing social contact under certain conditions¹⁰. The most common condition being the presentation of tasks that the child may find unpleasant and unrewarding and consequently they do not want to do them¹¹. This process is depicted in Box 5.6. The numbers in the figure refer to the sequence of events that are listed here:

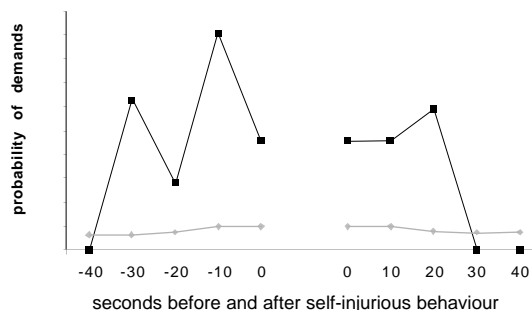
Box 5.6: Social reinforcement of self-injurious behaviour 2: Negative reinforcement (see main text for description)



1. The child is being asked to carry out a task which they do not want to do as they find it: difficult, hard work, painful, unrewarding, or any combination of these factors.
2. The self-injurious behaviour occurs. (Initially, the self-injury may occur at this time simply by chance or as part of a “tantrum” in which the child accidentally self-injures).
3. The other person finds the self-injury unpleasant or aversive and as a result of this does something in order to prevent another self-injurious response or tend to the results of the first response.
4. The other person engages with the child in order to prevent the self-injury. The response that the other person is making at this point may be no different to that described in the attention example given above. The important point here is that whilst engaging with the child the unpleasant task stops, at least temporarily.
5. The child finds this removal of the unpleasant task rewarding. This makes it more likely that the next time the child is presented with an unpleasant task (and they are thus motivated to escape the task) they will self-injure.

Box 5.7: Self-injury rewarded by escape from demands

Nathan was a 21-year-old man with Cornelia de Lange Syndrome. We visited him at his day centre and observed him for about four hours over the course of a typical day. Before we arrived we had been told by his parents that he sometimes engaged in self-injurious behaviour. When spending time with him it seemed that he would be more likely to self-injure when demands were made of him. The demands that were made mainly took the form of asking him to take part in particular activities or to move to other locations in the day centre. Following our visit we carried out a statistical analysis in which we looked at all the incidents of self-injury that Nathan showed. The graph below shows how just prior to his self-injurious incidents, the probability of demands occurring (black line) increased in comparison to the average level of demands over the day (grey line). Following the self-injury, the probability of demands quickly decreased. It is likely therefore, that his self-injury led to a withdrawal of demands by his carers and thus negatively reinforced (rewarded) the behaviour.



6. The child is being asked to carry out a task which they do not want to do as they find difficult, hard work, painful, unrewarding or any combination of these factors. (Go to 2 above).

As part of our research we observed a young man with Cornelia de Lange Syndrome who showed self-injury at his day centre. When tasks

Box 5.8: The ABCs of self-injurious behaviour

Antecedents: these are events or situations that occur immediately before any behaviour. In the examples we have just seen this may be being left unattended, (if the self-injurious behaviour is reinforced by adult attention) or being presented with a difficult task (if the behaviour is reinforced by escape from demand). Antecedents can be viewed as a trigger for the behaviour, just as being hungry triggers food seeking behaviour.

Behaviours: these are the behaviours shown by individuals (not parents or teachers) that evoke reinforcement. Challenging behaviours are the most commonly discussed as they tend to be very efficient at eliciting a response (and we are often studying them in order to try and decrease them!) but any verbal or physical behaviour shown by the individual comes into this category.

Consequences: these are the events, behaviours or sensations that immediately follow a behaviour. In the examples we have seen these may include physical attention (e.g. hugs), verbal reprimands, removal of a difficult task or the flashing lights seen by a child engaging in eye pressing. Consequences are usually reinforcing but if the consequence is not the usual reinforcer (i.e. the difficult task is not removed after SIB as usual), then the behaviour will often escalate in intensity until the reinforcer is forthcoming.

Example:

→A: child is unattended and has not had attention for some time.

→B: child engages in self-injurious behaviour.

→C: parent engages with child and provides attention.

occur when self-injury is rewarded by escape from demands (see Box 5.7).

In both these processes learning is taking place on the part of the child or adult. Each time the child is in this situation and shows the behaviour and is rewarded, this strengthens the association between the situation, the behaviour and the reward and makes it much more likely that this will occur in the future (see Box 5.8).

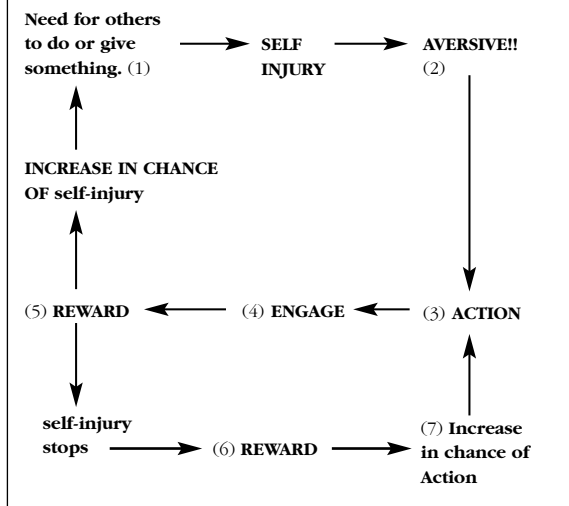
It should be noted that in both of these examples only one type of reward is being considered. In the positive reinforcement example, attention appears as the reward. However, it is entirely possible that along with the attention any number of other rewards are also present. This may include food, drinks and activities in order to try and distract the child from the self-injurious behaviour. When this happens the association between behaviour and reward is strengthened further as the child learns that self-injury leads both to attention and the presentation of these other items. Similarly, it may be the case that it is not necessarily just unpleasant task demands that are removed after self-injurious behaviour. It is possible that for children who do not like social contact that self-injury can be reinforced simply by the removal of social contact that does not have any demands associated with it¹².

These descriptions of the process of rewarding self-injurious behaviour have only focussed on the way in which the child is rewarded for showing self-injurious behaviour. It is important to also think about the other person in this interaction and how their behaviour is also rewarded⁸. If we consider this process within the positive reinforcement example that we have looked at above, then we can see that not only is the other person rewarding the child but the child is also rewarding the other person. This process is shown in Box 5.9. The numbers in the figure in Box 5.9 refer to the sequence of events described here:

1. The child is on their own and has no stimulation. Initially, the child may show self-injurious behaviour either because it gives rise to

were presented during the day or he was required to change activities this often led to a burst of self-injury and then the demand was removed. This is the chain of events that tends to

Box 5.9: Mutual reinforcement of self-injurious behaviour (see main text for description)



- pleasant sensory stimulation or because it relieves discomfort or as the end result of a stereotyped behaviour or it is simply a chance act.
2. The self-injury occurs and seen by another person (parent or carer).
 3. The other person finds the self-injury unpleasant or aversive and consequently acts to stop the self-injury from recurring or tries to find out the reason for the self-injury.
 4. The other person engages with the child who has just shown self-injurious behaviour and whilst preventing further instances of self-injury and trying to find the cause may comfort, distract or restrain the person or use any combination of these strategies.
 5. As the child has now received a reward there is no longer any motivation for the self-injury to continue and the self-injurious behaviour stops.
 6. As the self-injurious behaviour has now stopped the other person has been rewarded by the removal of the unpleasant event (the self-injurious behaviour). This reward is the feeling of relief that happens when a child stops self-injuring, even for a short period of time.

7. As the response by the other person to the self-injury is rewarded it makes it more likely that the person will make the same response to the self-injurious behaviour in the future and so reward the person again.

It is worth thinking about some other things that are always occurring in this process. One is what happens if the other person does not make a rewarding response to the self-injury. Under these circumstances the child will still have a need for the reward (i.e. is motivated see step 1 in Box 5.9) and so the self-injury will continue. As the self-injury continues so the other person will present more things until they eventually hit on the right thing and then the child will stop the self-injury. In this way the person is inadvertently taught by the child precisely how to reward the self-injury.

This sequence of events shows that just as the other person is inadvertently rewarding the child for showing self-injury, so the child is inadvertently rewarding the other person for rewarding the self-injury. This does not mean that the child intends to control the behaviour of the other person, it is simply the consequence of the natural sequence of events that occurs around any bout of socially reinforced self-injurious behaviour.

5.4: Self-injury as communication

When the social reinforcement process is described in this way it has led some people to describe self-injurious behaviour as being very similar to communication^{13, 14}. That is the self-injury is able to affect the behaviour of other people in the same way that communication can. With self-injurious behaviour that is socially reinforced the child effectively has the capacity to ask for things and refuse things. This is a useful analogy as it allows us to understand that self-injurious behaviour can in fact be a very adaptive behaviour in some respects. This is particularly the case when people have poor expressive communication and a limited repertoire of behaviours that they may call upon in order to

affect the behaviour of others. It was noted in a Chapter 3 that poor expressive communication and a greater degree of intellectual disability are risk factors for development of self-injurious behaviour. When the mutual social reinforcement process is operative, self-injurious behaviour can come to substitute for a limited expressive communicative ability.

There are two points which should be made with regard to the similarities between self-injurious behaviour and communication. The first is that the analogy only stretches as far as the pragmatics of communication i.e. the capacity for behaviour to influence the behaviour of others. Unlike language there are few rules and as a communicative act the behaviour is extremely crude. Second, when the analogy is applied to the process that is described in Box 5.8 it is clear that self-injurious behaviour is not necessarily due to a frustration with communication. Rather, it is a communicative act in itself.

It is worth thinking about the communicative analogy with regard to some specific features of Cornelia de Lange Syndrome. First, in Chapter 2 we showed that children and adults who have Cornelia de Lange Syndrome have very poor expressive communication and we had previously identified poor expressive communication as a risk factor for self-injurious behaviour. Thus, this risk marker can interact with the reinforcement process to cultivate self-injurious behaviour. Second, as we described in Chapter 2, children and adults with Cornelia de Lange Syndrome tend to have a severe or profound degree of disability. By definition this means that they have a limited repertoire of behaviours with which to influence others. Under these two circumstances self-injurious behaviour that is socially reinforced in a way that makes it communicative can emerge and easily become established in an individual's repertoire.

5.5: The effect of "setting events" on self-injury

Within the last ten years more attention has been paid to the fact that even when self-injurious behaviour is socially reinforced it tends to vary in

frequency across days and can also vary within a day. If the social reinforcement theory was a sufficient explanation then this should not necessarily be the case. In order to account for this variability researchers have looked at what are called "setting events" to try and understand why self-injurious behaviour should be occurring frequently at some times and not at others. The term "setting event" refers to something that influences the relationship between a situation and the likelihood that a behaviour will be shown.

One type of setting event that is relevant to understanding self-injurious behaviour in children and adults with Cornelia de Lange Syndrome is pain and discomfort. As we have discussed there have been a number of demonstrations that, for example, self-injurious behaviour can be worse when demands are made on individuals and in the past the self-injury has been reinforced by the removal of demands (see Box 5.6)¹⁵. It has also been shown that this relationship may be even stronger when particular setting events such as low mood, fatigue or illness are present^{16, 17, 18}. In other words demands may be made at times when people are not experiencing low mood, fatigue or discomfort and this will not lead to self-injurious behaviour. However, on other occasions if an individual is experiencing low mood, is tired or is experiencing discomfort and then a task demand may trigger the self-injurious behaviour. The reason for this is that the motivation for escaping a task is usually higher when anyone is tired or in discomfort (do you want to clean the house when ill or tired?) This shows how some biological factors might interact with psychological factors to raise the probability that a behaviour will occur.

5.6: Changes in the severity of self-injury over time

Whilst the social reinforcement theory can account for why self-injurious behaviour may be maintained or may continue, it cannot necessarily explain why the self-injurious behaviour occurred in the first place. For the social rein-

forcement theory to be right the behaviour has to occur before it can become reinforced. To explain this problem it has been suggested that self-injurious behaviour that is socially reinforced first started for another reason before becoming rewarded either by social contact or the removal of demands. The idea is that the behaviour may occur because it is reinforced by the sensory stimulation (see Section 5.3.1) or in response to a minor illness or discomfort or as a chance act, for example during a tantrum. Once the behaviour has occurred under these circumstances it may then become reinforced in the way that is described in Sections 5.3.2 and 5.3.3.

Once self-injurious behaviour becomes social-

Box 5.10: More reward for more severe self-injurious behaviour

The way that others respond to an individual's self-injurious behaviour can shape the form the behaviour takes and even the frequency and intensity of the behaviour. Here's how. Imagine your child is in the same room as you but you are not attending to them. Your child then proceeds to hit their head on the soft cushion of the sofa several times. How would you respond to this? Now imagine that instead of the soft sofa cushion, your child hits their head on the sharp corner of the coffee table. How would your response to this behaviour be different? Chances are you would respond to this second, more severe self-injury more quickly and more frequently (i.e. every time it occurred). This makes this form of self-injury more efficient for the child as it is going to result in reinforcement (in this example, in the form of parent attention) very quickly, each time it happens. This makes it more likely that this behaviour will be chosen over a less intense behaviour (head banging on the cushion) for which the likelihood of reinforcement from the parent is lower. As parents get used to a particular behaviour and start to ignore it, the child will progress to a more intense behaviour in order to return to the same level of efficiency as before, leading to an ongoing escalation in the severity of self-injury.

ly reinforced and occurs regularly there are some reasons why it may gradually become severe over time. In order to understand this increasing severity over time it is important to remember the reason that people reward (reinforce) self-injurious behaviour. Self-injurious behaviour is reinforced by other people because they experience it as unpleasant or aversive and want to stop it. It then follows from this that when a behaviour is more severe or potentially injurious then people are more highly motivated to stop the behaviour and thus more likely to respond and reinforce the behaviour. (see Box 5.10). When we consider the development of self-injurious behaviour over time it becomes easy to see why self-injury might increase in severity. It is highly likely that it is the more injurious or damaging responses that lead to a much quicker rewarding response by others. Consequently the child learns to show a more damaging response as opposed to a less damaging one. Gradually over time this ratcheting up of severity will lead to more damaging behaviour.

There is another way in which self-injurious behaviour might increase in severity when it is socially reinforced. When any behaviour is reinforced or rewarded it is possible to decrease the behaviour by simply withholding the reward or reinforcement. This is called extinction. However, the behaviour does not necessarily decrease immediately. In fact it shows a characteristic pattern which is called an extinction burst¹⁹. This means that the behaviour increases in frequency and intensity before finally decreasing. If when a behaviour such as self-injury is being socially reinforced an attempt is made to withhold the rewards, this is usually done by ignoring the behaviour. When this happens the behaviour may increase to such an intensity that it is impossible not to respond and consequently reinforce the behaviour. When this happens the person has then learned not to show the behaviour at a low frequency and low intensity but to show a much higher intensity and frequency of the behaviour. In the future the behaviour will then occur at this high rate and intensity.

The reinforcement or greater reward for more severe self-injurious behaviour is best understood by thinking about what is called the response efficiency of the behaviour²⁰. When a behaviour is socially reinforced the severity of the behaviour is to some extent determined by its efficiency. A behaviour that is very efficient will have very little cost to the individual, will take little effort to carry out and will result in no pain or discomfort but will have a very high return. That is, it will commonly be reinforced on every occasion, with a great deal of reward and this will occur very quickly. Under these circumstances we would say that a behaviour has high response efficiency. For self-injurious behaviour, one of the balances to the reinforcement that occurs is the potential pain or discomfort that accompanies each and every act of self-injurious behaviour. In other words, if the reward for showing self-injury was very small but the pain was very high then the behaviour would tend to occur very infrequently or not at all because the cost of the behaviour outweighs the return. However, it is important to remember that pain, particularly pain in the peripheral nervous system, maybe dampened in individuals with Cornelia de Lange Syndrome (see Chapter 2) and this may mean that there is a lower cost to the behaviour than would otherwise be the case.

5.7: Loss of control

In Chapter 3 it was noted that some people who shows self-injurious behaviour also show behaviours that seem to indicate that they are unable to control their own self-injury. These behaviours were referred to as self-restraint and a preference for imposed restraint. It is difficult to understand why behaviours such as self-restraint should occur and why people should want to be restrained if the self-injurious behaviour is acting as a form of communication. The social reinforcement theory suggests that the behaviour is under the control of the individual and the behaviour is shown when particular motivational conditions arise. If this is the case then it is diffi-

cult to see why someone should try to actively restrict their behaviour by seeking restraint when they could just simply not show the behaviour.

The argument that is often put forward in response to this is that the self-injurious behaviour is painful and consequently the person does not want to experience the pain²¹. However, if this is the case then it is not clear why the person does not simply stop showing the behaviour because the response efficiency is now imbalanced and there is now a comparatively poor return for the behaviour. Consequently, when self-restraint is occurring it seems likely that the social reinforcement theory might be less applicable.

Under these circumstances it has been suggested that the self-injurious behaviour has taken on a “compulsive” quality and consequently the individual will feel driven to show the self-injurious behaviour or may not be able to inhibit the behaviour²². The self-restraint then arises as a method of self control of a behaviour that the individual experiences as painful but is unable to inhibit. In Chapter 4 we described a study in which we had found that for those individuals with Cornelia de Lange Syndrome who showed self-injurious behaviour and other compulsive behaviours they were more likely to show self-restraint and appeared to be trying to exert control over their own self-injurious behaviour. Clinically, we have often observed self-restraint in people with Cornelia de Lange Syndrome who show self-injurious behaviour and it is a real possibility that for these individuals the behaviour is not completely under their control and they are trying to seek some external help in managing the behaviour. Their solution to this problem is self-restraint.

It is interesting to speculate why this problem might arise more in Cornelia de Lange Syndrome than we might expect. The answer might lie in disturbance of the serotonergic system. In Chapter 2 we noted that compulsive behaviours are much more common in individuals with Cornelia de Lange Syndrome than a comparable group of individuals with the same degree of intellectual disability. Additionally, a study in the

1970s showed that serotonin levels in Cornelia de Lange Syndrome were lower than normal²³. We have previously noted that one potential cause of compulsive behaviours is serotonergic disturbance. Although speculative at this stage it is entirely possible that a fundamental disturbance of serotonin exists in children and adults with Cornelia de Lange Syndrome and that when self-injurious behaviour occurs it becomes compulsive and consequently the individuals are unable to control their behaviour.

5.8: Summary

It is likely that the causes of self-injurious behaviour in children and adults with Cornelia de Lange Syndrome are not very different from other people who have an intellectual disability. Medical conditions associated with pain and discomfort, self-injury rewarded by sensory stimulation and the presentation or removal of social contact are all reported as important causes in all people who have intellectual disability. The difference for people with Cornelia de Lange Syndrome may well be that: they experience more medical conditions that can give rise to self-injury, they have poorer expressive communication, consequently self-injury may be more likely to come to serve a communicative purpose, and that they may be more likely to experience difficulties in inhibiting a self-injurious response than other people.

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Chapter 6: Assessment of the causes of self-injurious behaviour

6.1: Introduction

When starting to assess the causes of self-injurious behaviour in people who have Cornelia de Lange Syndrome, it is important to remember that for any child or adult self-injurious behaviour has not always occurred, and right now it does not occur all of the time (it just feels like that sometimes!). This means that the self-injurious behaviour is likely to be related to either internal or external factors that have changed over a long period of time or change on a more short-term basis. Successful assessment of the causes of self-injurious behaviour rests on finding out more about the factors that appear to be present when self-injurious behaviour is occurring, and absent when it is not.

In order to uncover the causes of self-injurious behaviour it is important to adopt a systematic approach to looking for the factors that appear to be associated with the behaviour. To develop this systematic approach we have drawn on two areas of knowledge. First, we have considered relevant factors that might be specific, or at least more common, in children and adults with Cornelia de Lange Syndrome. Second, we have drawn on the general principles of assessing self-injurious behaviour in anyone who has an intellectual disability. In combination, we believe an approach based on these two bodies of knowledge will give us the best chance of understanding self-injurious behaviour in any individual with Cornelia de Lange Syndrome.

In this chapter we will describe a systematic approach to understanding factors that may be causing self-injurious behaviour. Some of the assessments can be carried out by anybody who

has a good knowledge of the person showing self-injurious behaviour. However, some assessments require more expertise and the reader is encouraged to seek collaboration and co-operation with the right professionals. We do believe it is always better to seek the support of others when conducting these assessments in order to ensure that the approach is both systematic and thorough.

This chapter is broadly divided into three main areas. First, we will consider assessment of internal factors that can be related to self-injurious behaviour. Second, we will consider assessment of external factors that might explain self-injurious behaviour and third, we will consider assessment of factors that might account indicate a “loss of control” over self-injurious behaviour. It is important when thinking about assessing self-injurious behaviour to remember that the cause of self-injurious behaviour may not be related to a single factor and that a number of internal and external factors might be influential. It is also important to remember that causes can change over time in any individual and consequently it is important to repeat the assessment process at various points.

6.2: Assessing the role of pain and discomfort in self-injury

In Chapter 2 we noted that children and adults with Cornelia de Lange Syndrome may experience a number of short-term or long-term medical problems that can give rise to physical discomfort and pain at various times throughout their lives. We also noted that when people experience pain and discomfort they naturally try

to relieve the discomfort, usually by physically stimulating the area of the body that is associated with discomfort. In particular, we noted that children and adults with Cornelia de Lange Syndrome can experience discomfort around their eyes (due to lack of tear production and eyelashes touching the eye), the ears (with middle ear infections), the sinuses, the teeth (either because of crowding of the teeth, the increased risk of tooth decay due to thin enamel or the effect of reflux), gastrointestinal pain in the upper chest and throat (due to the stomach acid irritating the lining of the digestive tract) and, possibly, joint pain. Any of these medical problems can give rise to localised pain or more general discomfort and this can lead to a self-injurious behaviour being focused on or around the site of the pain or discomfort.

In order to assess whether any of these medical reasons might be associated with self-injurious behaviour it is important to assess the following factors:

Box 6.1: Eye problems and self-injury in Cornelia de Lange syndrome

At one Cornelia de Lange Syndrome conference we met a 32 year old man with self-injurious behaviour that mainly comprised punching his eyes and the area around his eyes. In fact, the self-injury was so severe that he was accompanied by two carers who restricted the movement of his arms so that the punching was not too damaging. His self-injury did not seem to be related to environmental events and had recently worsened. The ophthalmologist attending the conference examined his eyes and found that the eyelids had turned slightly inwards so that the eyelashes were now touching his eyes. The carers described a period during which the man's eyes had become dry and itchy and at this time he rubbed his eyes. It is possible that this made the eyelids swell and thus the eyelashes come into contact with the eye.

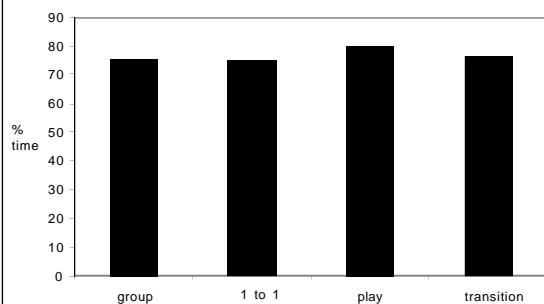
The site of the injury. Is the self-injurious behaviour directed towards any specific site of the body that is associated with a medical condition that we know is more common in children and adults with Cornelia de Lange Syndrome? As we have noted above, this means is the self-injury directed towards the eyes (see Box 6.1), ears, cheeks and bridge of the nose (where the sinuses are), the teeth or mouth, the chest or throat (where the discomfort associated with a reflux would be found).

The association between self-injury and environmental events. Self-injurious behaviour that is associated with pain and discomfort is unlikely to change as events in the environment change. This is because the cause of the pain or discomfort is highly unlikely to be related to environmental events. So, for example, the self-injurious behaviour will not be triggered by a change in the amount of social contact that an individual is experiencing, the person being asked to conduct a task or any other events that are occurring throughout the day (see Box 6.2). However, it may be the case that self-injurious behaviour that occurs more commonly after mealtimes than before them, is associated with gastrointestinal problems. Keeping a record of the times in a day that self-injurious behaviour occurs might reveal a pattern that shows that self-injurious behaviour is related to meal times in this way (see Section 6.3.4 on Scatterplots).

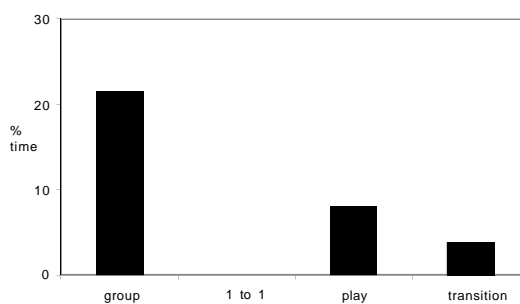
Change in other behaviours around the time of self-injury. The third aspect of assessing whether self-injurious behaviour is associated with pain and discomfort is to look for other indicators of discomfort that appear to be present when the self-injurious behaviour is more common. For someone who has poor communication it is often difficult to know whether the person is experiencing pain and discomfort. Some of the more obvious signs that someone is experiencing discomfort may be facial expression and vocalisations, loss of appetite, poor or disrupted sleep and a generally higher level of movement. Any or

Box 6.2: Self-injurious behaviour, pain and discomfort and environmental events

The graphs below show the percentage of time two children with Cornelia de Lange Syndrome spent showing self-injury in four different environmental settings.



1) For this 15 year old boy the self-injury consists of picking and scratching the area around the chest and this suggests the self-injury may be related to discomfort from reflux. As the graph shows, the self-injurious behaviour occurs at very high rates across all environmental settings indicating that the behaviour is not related to environmental changes. In combination these factors suggested that the self-injurious behaviour may be related to pain or discomfort. Following medical investigation for gastrointestinal problems, the self-injury decreased significantly.



2) For this 6 year old girl the self-injury comprised skin picking and scratching. The graph shows that the rate of self-injurious behaviour is very variable across different environments. The behaviour is very high in the group setting and completely stops in periods of one to one attention. This indicates that the behaviour is related to environmental settings. In this case the behaviour appears to occur more often in situations where attention received by the individual is likely to be low, but stops when attention is increased. We think that this behaviour might be rewarded by attention.

all of these indicators when seen with self-injurious behaviour might indicate that pain or discomfort is an underlying cause for the self-injury. (Behaviours that we believe indicate gastrointestinal problems are described in Chapter 2).

It is, of course, extremely difficult to ever be certain that medical reasons are associated with self-injurious behaviour, particularly when the person is unable to communicate. However, the three indicators listed above would suggest that medical reasons should be explored prior to any other reasons. If more than one indicator is present then we would suggest that a possible medical reason is likely.

6.3: Assessing reward by stimulation or the presentation or removal of social contact

When thinking about reasons for self-injurious

behaviour it is important to remember the information that was presented in Chapter 5 in which we discussed how self-injurious behaviour might be rewarded or reinforced by sensory stimulation (an internal factor) or social processes (an external factor). When beginning an assessment of possible psychological reasons it is usual to try to distinguish between self-injurious behaviour that might occur for sensory stimulation and that which might occur because of social processes. This is because the types of intervention that would be used are very different for self-stimulatory and socially reinforced self-injurious behaviour.

Before describing the main assessment strategies that can be used to evaluate these reasons for self-injurious behaviour it is important to note a number of issues that always need to be considered. First, there are always risks and benefits

associated with some psychological assessments. Risk of injury whilst assessing is a possibility for some of the experimental methods of assessment which often require the creation of conditions under which self-injurious behaviour usually occurs to ensure that we are right about a particular cause. Under these circumstances the risks to the individual from showing self-injurious behaviour and the benefits of being more certain about the causes of self-injurious behaviour need to be balanced and thought through with others.

It is also important to note that when we assess psychological reasons for self-injurious behaviour it is unlikely that we will be able to see a one-to-one relationship between self-injury and internal or external factors. It is more likely that we will just see a higher level of self-injurious behaviour when a cause is present compared to when it is absent, as opposed to being able to turn self-injurious behaviour on and off completely. It is also important that we think about different factors in different environments¹. This is most commonly the case between school and home environments. The things that happen in school may be very different to those that happen at home and the way that teachers and parents might respond to self-injurious behaviour will also differ. When considering the reasons for self-injurious behaviour it is important to consider differences between the main environments in which the child or adult might spend their time. Just as there are differences between environments that might be related to the reasons for self-injury, so the reasons can change over time. The reason for self-injurious behaviour to start in a young child might be very different from the reason that the behaviour continues over a longer period of time. The most common example of this is for self-injurious behaviour to begin for a medical reason or because of the stimulation that is produced by the self-injurious behaviour, and for the behaviour to then become socially reinforced which may maintain the behaviour over long periods of time (see Chapter 5)^{2,3}. For this reason it is important to remember that assessments for psychological reasons

should be repeated periodically.

One of the most important aspects of assessment is systematic record keeping and the value of good records cannot be underestimated. Whatever assessment is conducted the results must be carefully documented so that a comparison can be made at different points in time or between environments. It does not matter that an assessment does not show a positive result i.e. that a factor cannot be said to be associated with self-injurious behaviour. The results of this negative finding must still be documented so that others can see the results at a later date and make a comparison should that be necessary. The key to conducting a good assessment is good record-keeping. We outline some methods of keeping records in Chapter 7.

There is a very large research literature covering the assessment of psychological reasons for self-injurious behaviour. Basically this literature identifies five different reasons that self-injurious behaviour may occur and these different reasons are directly related to the causes of self-injurious behaviour that are described in Chapter 5. These five reasons are:

1. Sensory reinforcement or stimulation (an internal factor).
2. Positive social reinforcement, most commonly by attention. (an external factor).
3. Positive tangible reinforcement by the presentation of things or activities such as food, drink or toys etc.. (an external factor).
4. Negative social reinforcement by escape from tasks or activities that the person does not want to do. (an external factor).
5. Negative social reinforcement by escape from any social contact regardless of whether or not tasks or activities are involved. (an external factor).

When psychologists undertake the assessment of self-injurious behaviour they are trying to find out which of these reasons apply to the self-injury so that they can match the treatment to the reason. The assessment task therefore is to

systematically evaluate each of these potential reasons.

There are a number of ways in which these reasons can be assessed that range from informal information gathering, by asking those who know the child or adult well and conducting informal observations, through to conducting experimental methods in which environmental events are systematically manipulated and the effect on self-injurious behaviour is recorded. In addition to these assessments of self-injurious behaviour there are some additional assessments which are important to conduct in order to help develop the best intervention and we will consider these towards the end of the chapter.

6.3.1: Informal information gathering

The benefits of this type of assessment are that it is quick to conduct and that it can cover a wide range of environments and events. However, a problem is that the information that is collected may be unreliable, and there is good evidence that if you ask different people about the events that are associated with self-injurious behaviour for any individual then you tend to get different answers. For this reason psychologists would usually use this method to collect some basic information and then would go on to use one of the other methods that have been described below.

Basically, when conducting this kind of assessment you are trying to identify the events that happen before self-injurious behaviour occurs (these are called antecedents) and the events that happen after self-injurious behaviour has occurred (these are called consequences). Thus, the overall analysis is called antecedent-behaviour-consequence or ABC for short and we outlined this idea in Box 5.7.

The main strategy in this assessment is to ask about different antecedents that appear to be occurring before self-injurious behaviour and that would indicate one of the five reasons that are described above. So, if self-injurious behaviour tends to occur more when there is a boring

and unstimulating environment than when there are things to do, then it is likely that the self-injurious behaviour is maintained by sensory stimulation (and consequently the model outlined in a Box 5.4 is likely to be applicable). If self-injurious behaviour tends to occur when a carer is not attending to the person who shows self-injury and is attending to someone else, then it is likely that the self-injurious behaviour is maintained by a positive social reinforcement (and consequently the model outlined in a Box 5.5 is likely to be applicable). If self-injurious behaviour occurs more when the person is asked to do something they do not want to then it is likely that the self-injurious behaviour is maintained by negative social reinforcement of escape from task (and consequently the model outlined in Box 5.6 is likely to be applicable). The questions that might be asked and the informal observations that might be conducted in order to find out which of the five reasons outlined above are likely to be applicable are described in Box 6.3.

6.3.2: ABC and STAR charts

There is really very little difference between an ABC chart and a STAR chart and both seek to obtain a record of what happens before and after the self-injurious behaviour. STAR stands for Setting, Trigger, Action, Results and so gives a bit more information than ABC charts in that you learn about the settings in which the behaviour is likely to occur^d. ABC and STAR charts are commonly used by psychologists to conduct the assessment of psychological reasons for self-injurious behaviour when the behaviour is occurring at a relatively low frequency, say a couple of times a day. If the behaviour is much more frequent then ABC and STAR charts tend to be less informative and the observational or experimental methods that are discussed below might be more appropriate.

There are two types of ABC charts that might be used for assessing self-injury. Open ended ABC charts simply require people to record in boxes the events that they saw just before the

Box 6.3: Assessing the function of self-injurious behaviour

Self-stimulation: When considering whether self-stimulation may be the function of a given form of self-injurious behaviour you may be asked to indicate whether your child or person you care for:

- ❖ Does this because he/she is bored?
- ❖ Does this most when left alone or thinks they are alone?
- ❖ Seems to do this simply for the pleasure/sensation it provides?
- ❖ Does it repeatedly for extended periods, even if they have access to toys?

Positive social reinforcement: When considering whether positive social reinforcement may be the function of a given form of self-injurious behaviour you may be asked to indicate whether your child or person you care for:

- ❖ Does it to get a response from an adult?
- ❖ Does it to try and attract your attention?
- ❖ Does it to annoy you and make you reprimand them?
- ❖ Does it so that you will come and talk or play a game with them?
- ❖ Does it when you are talking to someone else in the room?

Positive tangible reinforcement: When considering whether positive tangible reinforcement may be the function of a given form of self-injurious behaviour you may be asked to indicate whether your child or person you care for:

- ❖ Does it because they're hungry or thirsty and want you to get them something?
- ❖ Does it if you take their favourite toy away from them?
- ❖ Does it if a sibling has a toy they want to play with themselves?
- ❖ Does it if a sibling takes something from them?
- ❖ Does it as a way of asking someone to fetch something for them?

Negative social reinforcement: When considering whether negative social reinforcement (escape from either social contact or demands) may be the function of a given form of self-injurious behaviour you may be asked to indicate whether your child or person you care for:

- ❖ Does this when asked to do something (i.e. tidy room, wash face, school work etc)?
- ❖ Does this when asked to go somewhere they don't like (i.e. dentist etc)?
- ❖ Does it as a way of getting someone to leave them alone?
- ❖ Does it when they don't want to join in with something?

self-injury occurred (A, antecedent), a brief description of the behaviour itself (B, behaviour) and a description of the events that happened just after the behaviour (C, consequence). An example of a completed ABC chart is shown in the Box 6.4. together with an interpretation of how the completed form is related to the five causes.

A second way in which ABC or STAR charts may be used is by designing a closed-ended form that is completed for each incident or burst of self-injurious behaviour⁵. Using this method it is possible to help people who are keeping the records to keep an eye out for the types of event that would help to understand the possible rea-

Box 6.4: Using ABC charts to assess self-injury

	Antecedent	Behaviour	Consequence
1	Child is alone	Child presses on eye	Nothing
2	Parent is talking to a friend	Child bangs head on the floor	Parent stops talking and picks up child
3	Parent asks child to put away toys	Child bites hand	Parent takes child to another room
4	Parent is on telephone	Child slaps face repeatedly	Parent puts phone down and distracts child with biscuit
5	Parent and child are folding clothes together	Child bites fingers	Parent goes into another room
6	Parent and child are folding clothes together	Child bites fingers	Parent takes child into another room
7	Child is playing on own	Child hits head	Nothing
8	Parent and child are playing together	Child slaps face	Parent distracts child with favourite toy
9	Parent is playing with child's sibling	Child bangs head	Parent encourages child to join in
10	Parent is washing child's face	Child bites hand	Parent goes to another room

Items 1 and 7 are examples of sensory reinforcement. Although there is no obvious consequence in terms of behaviour by another person, the behaviour is reinforcing on its own so the stimulation the behaviour provides is the reinforcer.

Items 2 and 9 are examples of positive social reinforcement as the self-injurious behaviour results in attention from the parent and there was no attention before the self-injury.

Items 3 and 6 are examples of negative reinforcement by escape from a demand as the self-injury results in the demand being removed (in these examples by the child escaping the room altogether).

Items 4 and 8 are examples of positive reinforcement by access to tangibles as self-injury results in access to food (4) or a preferred item (8).

Items 5 and 10 are examples of negative reinforcement by avoidance of social contact as self-injurious behaviour results in the adult moving away from the child.

sons for self-injurious behaviour. An example of a closed ended ABC chart is shown in Box 6.5. Research that we have conducted in the past in which we have compared the information that can be taken from open ended and closed ended ABC charts strongly suggests that the closed ended ABC charts give better information, probably because they help the person completing the charts to look for the right things.

When reviewing the completed ABC and STAR charts, you are looking for antecedents and consequences that might indicate which of the five causes appear to be important. For example, if the records show a pattern of low levels of attention and carers being with other people and then the consequences show that some form of attention was paid following the behaviour then it is likely that the behaviour is maintained by

Box 6.5: Using a closed ended form to assess self-injurious behaviour

Below is an example of a completed closed ended form. Each time an inappropriate behaviour occurs all relevant items are marked.

What happened before episode?	Inappropriate behaviour	What happened after episode?
<input type="checkbox"/> The individual was on their own <input type="checkbox"/> They were asked to leave the table <input checked="" type="checkbox"/> Asked to wash the dishes <input type="checkbox"/> Someone took a toy from them <input type="checkbox"/> Someone was playing a game with them <input type="checkbox"/> They were asked to stop doing something	<input checked="" type="checkbox"/> Self-injurious behaviour	<input type="checkbox"/> Given a biscuit <input checked="" type="checkbox"/> Sent to their bedroom <input type="checkbox"/> Played with toys on their own <input type="checkbox"/> Everyone ignored the behavior <input type="checkbox"/> They were told off for behaviour <input type="checkbox"/> Parents gave them a hug

The example above runs as follows: immediately before the incident, the individual is asked to do something. Self-injury then occurs and the individual is sent to their room. This indicates that the behaviour may be maintained by negative reinforcement in the form of escape from task demand as the consequence of the behaviour is for the task to be removed (or in this case, the person is removed from the task).

positive social reinforcement (see Box 5.5). However, if the records show that prior to the behaviour occurring tasks are being presented or the person is being asked to do something and following the behaviour the task is no longer present, then it is likely that the behaviour is maintained by a negative social reinforcement by escape from a task (see Box 5.6). These are just two of the reasons that might be revealed by ABC and STAR charts and boxes 6.4 and 6.5 shows the sorts of antecedents and consequences that might be associated with the different reasons for self-injurious behaviour.

6.3.3: Questionnaire methods

There are two questionnaires that can be used to assess the factors that might influence self-injurious behaviour. They are the Motivation Assessment Scale (MAS)⁶ and the Questionnaire About Behavioral Function (QABF)⁷. These have been designed to ask specific questions about

possible factors. The person who is completing the form gives a rating for different questions and these ratings can then be added to give a score for different causes. Examples of the questions that are used in the questionnaires are given in Box 6.6 together with an interpretation of what the responses might mean. Unfortunately these questionnaires are not easy to get hold of but Clinical Psychologists will usually be able to obtain them.

6.3.4: Scatterplots

Scatterplots are slightly different from ABC and STAR charts and are usually used to get an indication of the times of day that the self-injurious behaviour tends to occur⁸. The advantage of this method is that it is not very time consuming for those who are being required to complete the charts and the information can be used as a good baseline by which to compare the effects of an intervention that is put into place following

Box 6.6: Example questions from the MAS and QABF

The Motivational Assessment Scale⁶ and The Questions About Behavioral Function questionnaire⁷, include items that refer to four types of reinforcement: self-stimulation, escape from demand, social reinforcement (attention) and tangible reinforcement. They provide a quick and easy way for parents and practitioners to try and understand problem behaviour. Examples from both scales are shown below:

Self-Stimulation:

MAS: Would the behavior occur repeatedly, in the same way, for very long periods of time, if no one was around? (for example, rocking back and forth for over an hour.)

QABF: Engages in the behavior in a highly repetitive manner, ignoring his/her surroundings.

Negative social reinforcement:

MAS: Does the behavior occur following a request to perform a difficult task?

QABF: Engages in the behavior to escape work or learning situations.

Positive social reinforcement:

MAS: Does the behavior seem to occur in response to your talking to other persons in the room?

QABF: Engages in the behavior to get attention.

Positive tangible reinforcement:

MAS: Does the behavior ever occur to get a toy, food or activity that this person has been told he or she can't have?

QABF: Engages in the behavior to get access to items such as preferred toys, food or beverages.

assessment. Basically, a scatterplot consists of a grid in which a rating of the frequency or intensity of self-injurious behaviour is made for each, say, half-hour or hour of the day. A completed example is shown in Box 6.7.

When reviewing a completed scatterplot it is helpful if the ratings that are made throughout the plot can be related to a diary of events and activities that the individual was involved in. This will help to try and identify the relationship between events and activities and levels of self-injurious behaviour so that conclusions might be drawn about possible external factors that are related to the behaviour. So, if high level self-injurious behaviour recorded in the scatterplot appears to be related to times at which there are one-to-one teaching sessions it is possible that the self-injurious behaviour is occurring because it is socially negatively reinforced by escape from the tasks (see Box 5.6). If high levels of self-injurious behaviour are recorded in the scat-

terplot at times when there is little activity and the person is left to their own devices, then it is possible that the self-injurious behaviour is occurring because it provides stimulation for the person. An example of the interpretation of the information from scattered plots are given in Box 6.8.

While scatterplots can give some very useful general information about the times of day that self-injurious behaviour is occurring, successful interpretation of the information depends on how well the level of self-injurious behaviour can be tied to a diary of events and activities, and how accurately the diary of events and activities is described. Sometimes, although self-injurious behaviour can clearly be shown to be associated for example with group activities, it is difficult to know whether this tells us enough to work out the reason for the self-injurious behaviour. It could be that the individual is not receiving a great deal of stimulation in this setting, and con-

Box 6.7: Scatterplot for recording the frequency of self-injurious behaviour

	Mon	Tues	Wed	Thurs	Fri
9.00	X	X		X	X
9.30			X	X	
10.00					
10.30					
11.00	X	X	X		X
11.30			X		
12.00	X	X		X	X
12.30	X	X	X	X	
13.00			X		
13.30					
14.00			X		X
14.30				X	
15.00					
15.30	X	X	X		X
16.00			X	X	
16.30					

The above figure is an example of a completed scatter plot. Time of day has been segmented into half hours. An “X” is marked if the behaviour has been seen within the half hour interval. In this case most of the self-injurious behaviour occurs around meal or snack times i.e. between 9.00 -9.30 (just after breakfast), 11.00 – 11.30 (around the time of a mid morning snack), between 12.00 -1.00 (around lunch time) and between 3.30- 4.00 (around tea time). With this pattern of data we might want to investigate which activities take place at these times (are the activities associated with low attention, low stimulation or high demands) and whether pain and discomfort is caused by gastro-intestinal problems and thus a cause of the self-injurious behaviour.

sequently self-injures because of self-stimulation, or it could be that there is shared attention and that the self-injury is reinforced by social attention from the carer who is working with the group. Results of scatterplots therefore should be reviewed alongside other assessment information.

6.3.5: Probability plots

Probability plots are similar to scatterplots but may prove to be more accurate when trying to evaluate whether a particular antecedent is associated with self-injurious behaviour and thus which of the five causes is important. We should emphasise that this is a relatively new method that we are currently developing in order to aid the understanding of the environmental reasons for self-injurious behaviour and to date it has not been widely used⁹.

A probability plot is conducted by a drawing up a chart that has times recorded down the left-

Box 6.8: Interpreting the function of self-injurious behaviour from scatter plots

Activity		Mon	Tues	Wed	Thurs	Fri
Group work	9.00		X			
	9.30				X	
Play time	10.00			X		
1 to 1 work	10.30	X	X	X	X	X
	11.00	X	X	X	X	X
Group work	11.30					
	12.00	X				X
Lunch time	12.30				X	
	13.00					
Play time	13.30	X	X	X	X	X
1 to 1 work	14.00	X	X	X	X	X
	14.30	X				
Play time	15.00					
Group work	15.30				X	
	16.00	X		X		
	16.30					

In this scatter plot the activities of the day that correspond to each half hour interval are also noted. This individual shows high levels of self-injurious behaviour during one-to-one teaching sessions (80% of all sessions) in which demands made on the individual are increased. In other sessions the rate is lower: Group is 14%; Play is 40% and lunch is 10%. On the basis of this information we might then suggest that this individual’s self-injury is likely to have a demand escape function.

Box 6.9: Probability plot for self-injurious behaviour

	SIB	Receiving attention	Not receiving attention	Receiving demands	Not receiving demands
1	X		X		X
2	X		X		X
3	X		X		X
4	X		X		X
5	X		X		X
6	X		X		X
7		X			
8		X			
9		X			
10			X	X	
11			X	X	
12	X		X	X	
13	X		X		X
14	X		X		X
15	X		X		X

This probability plot is similar to the scatter plots in boxes 6.7 and 6.8 but more specific. Here the time is split into 1 minute intervals and the caregiver's behaviour is also recorded in this plot. The plot above indicates that self-injury occurs when the individual is not receiving attention from the caregiver. The behaviour stops for a brief period when attention is being received and starts again almost immediately as the attention is once again removed. In this case we would suggest that the behaviour is attention maintained. The probability plot is much more specific than the scatter plot and can indicate the function of the behaviour more accurately.

hand side and environmental events across the top. There is also a box to record self-injurious behaviour. An example of a probability plot is shown in Box 6.9. Once the probability plot has been developed the observations are conducted throughout the day across a number of days. The method of observation to complete the plot is called Momentary Time Sampling (see Box 7.3) and involves the person who is spending time

with the child or adult who is showing self-injurious behaviour making a record at given time intervals. In the past we have used ten-minute intervals although more frequent observations can be used.

To complete the records the carer does not need to watch the child or adult all of the time. Rather, they will wear a watch which will give a signal, say, every 10 minutes (this is usually called the countdown function on digital watches) and at that point the carer will look at the child or adult for about five seconds and make a record of which environmental event is occurring and whether or not self-injurious behaviour is occurring. If the carer is unable to make the observation precisely at the time at which the signal occurs then this does not matter. However, it is important for the carer to make the observation as soon as possible after the signal has occurred. The important thing is that the observation does not occur just because self-injury is happening.

Once a number of probability plots have been completed the information can be analysed to try and identify a pattern of association between self-injurious behaviour and the environmental events. This is a little complicated but is worth the effort. Basically, the analysis consists of working out the probability (or chance) that self-injurious behaviour would occur. This means dividing the number of times that self-injurious behaviour was observed by the total number of observations that were carried out. The result of this calculation gives us a probability value. For example, if self-injurious behaviour was observed on 58 occasions and the total number of observations was 325, then the probability of self-injurious behaviour is 58 divided by 325 or 0.18 (these are odds of about 5 to 1 against). We can then calculate whether the self-injurious behaviour is more likely to occur given that an antecedent event has occurred. This means only looking at the occasions in which an antecedent has occurred and then calculating the probability of self-injurious behaviour. For example, if the social event of tasks was recorded on 65 occasions and when we look at those 65 occasions we note that self-inju-

Box 6.10: Calculating the probability of self-injurious behaviour

We can calculate the likelihood of self-injury (or any behaviour) occurring both across entire observation periods and in relation to environmental events such as task conditions. The data presented in box 6.9 will be used as examples of how to calculate probabilities and demonstrate what they mean.

Unconditional probability: this is the likelihood of self-injury occurring across the whole period of assessment. In box 6.9 there are 15 separate sessions in which behaviours were observed. self-injury occurred in 10 of these sessions. This is an unconditional probability of 0.66 (10/15) and odds of 1.5 to 1 against.

Conditional probability: this is the likelihood that self-injury will occur given a particular antecedent, such as low adult attention. Instead of counting the number of times self-injury occurs overall, we now look at the number of times it occurs when the individual is not receiving attention. In our current example (box 6.9) there were 12 instances when the child was not receiving attention and self-injury occurred in 10 of them. This gives a conditional probability of 0.83 (10/12) which is odds of 1.2 to 1 against.

There were also 3 sessions in which the child received demands and self-injury occurred in 1 of them. This is a conditional probability of 0.33 (1/3) and odds of 3 to 1 against.

As you can see, the odds of self-injury occurring when attention is low are higher than the odds for demand conditions, suggesting that self-injury is related to low levels of attention and thus probably rewarded by the presentation of attention.

rious behaviour occurred on 30 occasions then we would divide 30 by 65 and this gives us a probability of 0.46 (these are odds of about 2 to 1 against). When we compare the two probabili-

ties we can see a difference (5 to 1 against is more of a long shot than 2 to 1 against).

We describe this by saying that the simple or unconditional probability of self-injurious behaviour is 0.18 (5 to 1 against) and the conditional probability of self-injurious behaviour given that tasks are occurring is 0.46 (2 to 1 against). We can conclude from this analysis that self-injurious behaviour is approximately two and a half times more likely when tasks are occurring (0.56 divided by 0.18 or 5 divided by 2). When we look at our five causes it then seems likely that the reason for self-injurious behaviour is negative reinforcement by escape from task demands. Examples of the calculations for this analysis are given in Box 6.10.

6.3.6: Unstructured natural observations

Unstructured natural observations are difficult to conduct and the information can be difficult to analyse and carers will need help and advice from a clinical psychologist or somebody familiar with behavioural techniques. Basically, the method consists of using either paper and pencil or a palmheld computer to record at each second or, for example within a ten second interval, the self-injurious behaviour and the environmental events that are observed^{10, 11}. It is then necessary to analyse the data by looking at the probability that self-injurious behaviour is associated with events that happen before or after the behaviour with the probability that self-injury will occur regardless of what is happening. Thus, the analysis is similar to the comparisons of unconditional and conditional probabilities described above. Further information about this method can be found in some of the texts on applied behaviour analysis listed in the Bibliography and we have included this information here for the sake of completeness.

6.3.7: Analogue or experimental conditions

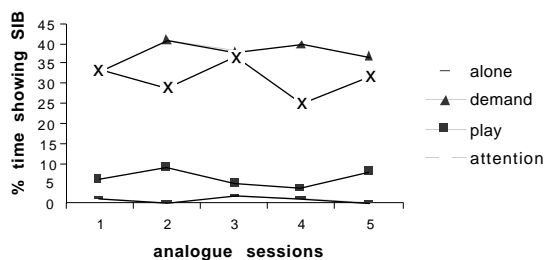
Over the last 20 years psychologists have developed a method of assessing the psychological reasons for self-injurious behaviour that

Box 6.11: Analogue analysis of self-injurious behaviour

Below we have presented the data from a functional analysis of self-injurious behaviour using analogue methodology. The numbers in the table represent the percentage of time self-injury occurred for in each of the conditions when we repeated each condition five times, once in each session.

	Session				
	1	2	3	4	5
Alone	1	0	2	1	0
Demand	33	41	38	40	37
Play	6	9	5	4	8
Attention	34	29	37	25	32

The graph below depicts the same data and shows clearly that SIB occurred at different levels in the different conditions. SIB occurred infrequently in the Alone and Play conditions but at much higher rates in the Attention and Demand conditions. This suggests that the self-injury in this case is likely to be maintained by both positive reinforcement in the form of attention and negative reinforcement in the form of escape from task demand.



involves systematically manipulating environmental conditions and then observing the effect on self-injurious behaviour¹². This methodology is usually referred to as applying analogue conditions (analogue meaning a model, in this case a model of what happens in the natural environment). The ideas behind analogue conditions are not themselves complicated but actually conducting the conditions can be difficult and it is important to use this method of assess-

ments in collaboration with a clinical psychologist or another professional who is familiar with behavioural methods.

We have already described the different psychological reasons that might be associated with self-injurious behaviour. In the observational methods we have described so far we rely on the natural occurrence of environmental conditions and self-injurious behaviour in order to see whether the self-injury occurs more frequently when specific environmental conditions are seen. Analogue conditions are designed as an experiment in which specific environmental conditions are presented and the frequency or duration of self-injurious behaviour is recorded. So, instead of waiting to see if low attention or high task demands trigger self-injury, these conditions are artificially created to see if they make self-injury more or less likely. There are two main types of analogue conditions that have been used in the past that can be employed to assess both the social and stimulatory reinforcement that form the basis to our five psychological reasons for self-injury.

Brian Iwata and his colleagues developed the first method in the early 1980s¹³. In this method the person who shows self-injurious behaviour is exposed to four different conditions a number of times and the conditions are presented in a random order. The conditions are:

Alone: The individual being assessed is left alone in a room with no toys or other forms of stimulation. No social consequences are delivered if self-injury occurs. If self-injury occurs more in this condition than others then it is probably maintained by self-stimulation.

Task demands: An experimenter (or teacher/parent) provides instructional tasks to the individual, using a three-step prompting system (verbal, gestural and physical prompts). The task is removed for several seconds if self-injury occurs. High levels of self-injury in this condition would suggest that the behaviour is maintained by negative reinforcement in the form of escape.

Social attention: The individual has access to toys. The experimenter provides several seconds of attention (i.e. saying “don’t do that”) following self-injury, but ignores the individual the rest of the time. High levels of self-injury in this condition would tell us that the behaviour is maintained by positive reinforcement in the form of attention.

Control (or play): The experimenter and individual play with toys as normal but no demands are delivered. No social consequences are delivered if self-injury occurs. This condition serves as a control condition to rule out the effect of other factors such as the experimenter being present.

Each condition lasts about ten minutes and throughout the condition a record is kept of the frequency or duration of self-injurious behaviour. Each condition is usually conducted about seven to ten times in order to insure that the results are stable. Once completed the results are graphed in order to examine which condition appears to cause a higher rate of the self-injurious behaviour. Box 6.11 shows an example of how the results of analogue conditions can be graphed in order to evaluate the possible psychological reason for self-injurious behaviour.

The second method was developed by Ted Carr and Mark Durand and was first described in 1985¹⁴. In this method the person who shows self-injurious behaviour is exposed to three different conditions in a systematic order. The conditions are:

High attention, easy task demand: In this condition, children carried out an easy task whilst receiving high levels of attention in the form of instructions, praise or neutral comments.

Low attention, easy task demand: In this condition, the task difficulty was the same as the baseline above but children only received attention in 33% of trials instead of 100%.

High attention, hard task demand: In this condi-

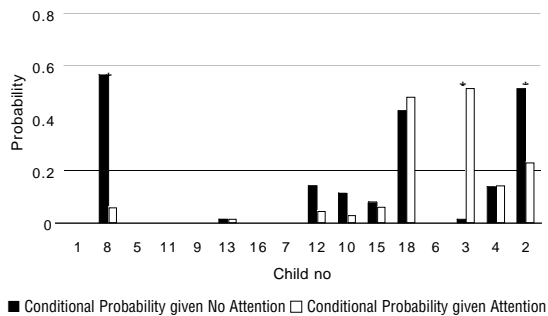
tion the child received attention on 100% of trials as in the baseline but the difficulty of the task was increased.

Again each condition lasts about ten minutes and a record of self-injurious behaviour is kept. Each condition is repeated approximately six times and the results graphed to examine which condition is associated with self-injurious behaviour. This method of analogue conditions only looks at the possible social reinforcement of self-injurious behaviour and not at whether the self-injurious behaviour occurs for self-stimulation. The interpretation of the results is carried out by comparing the rates of self-injury across the conditions. So, if the self-injurious behaviour occurs at much higher levels when there is low attention, easy task demands than when there is high attention and easy task demands it is likely that the self-injurious behaviour is occurring because of the low attention and consequently it is likely that the reason for self-injurious behaviour is to gain attention. However, if the self-injurious behaviour is occurring when there is high attention, difficult task demands in comparison to high attention, easy task demands then it is likely that the self-injurious behaviour is occurring to escape task demands (social negative reinforcement by escape from task demands.)

Whilst these two methods of analogue conditions are the most commonly used there are numerous variations that have been employed in order to try and understand the psychological reasons for self-injurious behaviour. Researchers have used so-called brief analogues¹⁵, with each condition lasting only five minutes in order to conduct the assessment more efficiently, or have used just two conditions to assess whether self-injury might be related to low attention (and thus socially reinforced by the presentation of attention). We used this method to look at the self-injurious behaviour of some children with Cornelia de Lange Syndrome who attended the Chester conference in 2001. Box 6.12 shows the results of our assessments.

Box 6.12: Analogue results from Chester conference in 2001

The graph below represents the results from a brief analogue study that was carried out with a group of children with Cornelia de Lange Syndrome at the Chester conference in 2001. In this study conditions of no attention and attention were alternated. Occurrence of self-injurious behaviour was recorded. The dark bars on the graph represent the probability or likelihood of self-injury occurring during sessions where no attention was given to the individual. The white bars represent the probability or likelihood of self-injury occurring during sessions where attention was given to the individual. A * above the graph represents a statistically significant difference. Two of the children who participated (2 and 8) were significantly more likely to self-injure during the no attention condition. We would therefore suggest that the function of self-injury for these children is to gain attention from others when it is not present. Only one child (3) was significantly more likely to self-injure during the attention condition. We might suggest that in this case self-injury was used to escape attention. For most children however, self-injury did not differ significantly between the conditions.



Additionally, researchers have explored some of the more idiosyncratic or unusual causes of self-injurious behaviour using analogue conditions. For example, for one child although generally difficult task demands did not necessarily cause self-injurious behaviour, when the demand

was that the child took part in a medical intervention this did cause high levels of self-injurious behaviour¹⁶. This is presumably because in the past medical interventions were associated with pain and discomfort for the child, whereas normal task demands were not, and the self-injury was reinforced by the medical examination being terminated. This brief example shows us two things. First, that we can use analogue conditions in order to understand specific causes for any given individual and second, that sometimes the normal analogue conditions that are used to may not reveal causes that are important for every child. These idiosyncratic causes might only be revealed in the informal interviews and observations that are described above.

6.4: The assessment of setting events

In a Chapter 5 we introduced the idea of setting events and illustrated how events such as periods of illness or fatigue can influence self-injurious behaviour by interacting with an antecedent. The most common examples that appear in the research literature are interactions between somebody being tired, unwell or experiencing low mood and the presentation of a demanding task. This is an example of how internal and external factors can interact. Under these circumstances the self-injurious behaviour may occur because the task is made so much more unpleasant by the fact that the person is tired or in discomfort.

The assessment of these potentially relevant setting events has been conducted in two ways. First, a record of the setting event (such as fatigue, low mood, being in pain or unwell) is kept using a diary or the scatterplot method. An additional record of the environmental events and self-injurious behaviour is also kept using the ABC or probability plot methods. The resultant records are then examined to see if the probability of self-injurious behaviour is higher when both the setting event and the antecedent are present as opposed to when either the antecedent or setting event is present. The sec-

ond method is to combine a diary record of setting events with analogue conditions. Using this method the effect of a given analogue condition is compared when the setting event is present with when it is absent. So, if the self-injurious behaviour only tends to occur when a person is experiencing in low mood and the analogue condition of task demands is presented, as opposed to when either of these alone is present, then this would indicate that there is an interaction between a setting event and an antecedent. An example of this analysis is shown in Box 6.13.

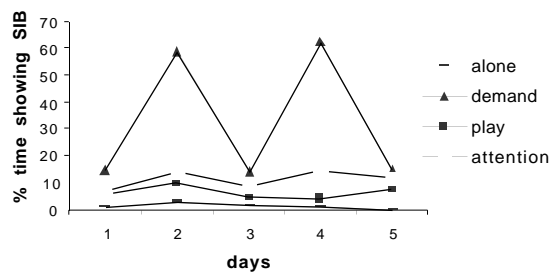
6.5: Assessment of communicative and adaptive behaviours and the broader environment

In addition to the assessment of self-injurious behaviour it is also important to assess a number of adaptive behaviours that might be shown by a child or adult in order to provide the groundwork for increasing behaviours that might replace self-injury. There are two areas of assessment requiring particular attention that are extremely important when self-injurious behaviour is occurring because it is reinforced by social events such as attention, more tangible things such as access to materials or food and drinks, and escape from demands or social interactions. These are communication and other adaptive behaviours that help the person exert control over their environment.

You will remember from Chapter 5 (Section 5.4) and the descriptions in boxes 5.5 and 5.6 that when self-injury is socially reinforced we can think of the behaviour as being very similar to communication. For this reason we must attend to the ability of an individual to communicate with others and by this we really mean the ability of an individual to affect the behaviour of others. This is called the pragmatics of communication. Additionally, we must also attend to the adaptive behaviours that an individual may already show or could show that would allow them to have control over their environment and thus satisfy their own needs. If the child or adult is unable to control aspects of their environment

Box 6.13: Interaction of setting events and self-injurious behaviour

Presented below are the data from an analogue analysis conducted over a period of five days. It shows the percentage duration of self-injury on each day. It shows that on two days, self-injury was highest during the demand condition, suggesting that the behaviour is maintained by escape from demands. However, it was also observed that the individual had had very poor sleep the night before testing on two occasions, days 2 and 4. The tiredness interacted with the demand condition to produce high rates of self-injury. These data show an interaction between a normal reinforcer and a setting event that leads to increased rates or a “double probability” of self-injury occurring.



directly then they will require others to help them to achieve this, and this makes it more likely that social contact is very rewarding. So, any assessment of self-injurious behaviour that is occurring because of social rewards should also attend to communication and other adaptive behaviour.

6.5.1: Communication

A speech and language therapist or a clinical psychologist should ideally conduct an assessment of the expressive communication of a child or adult with Cornelia de Lange Syndrome. When assessing for the purpose of building an intervention that might decrease self-injurious behaviour there are number of approaches to communication that might be adopted. The first is an assessment of the pragmatic communicative abil-

Box 6.14: Types of communication shown by children and adults with Cornelia de Lange Syndrome in our observational study of 54 children and adults

- ❖ **Vocalisations (not speech)**
- ❖ **Signing/Speaking**
- ❖ **Approach behaviours – moving towards another person/ reaching out to be lifted or hugged**
- ❖ **Touching/tapping/tugging another person**
- ❖ **Dissent behaviours – pushes adult's/ caregiver's hand/ body away/ wriggling out of physical contact/ moving away from the adult/ caregiver who is interacting with them.**
- ❖ **"Autistic" leading – using another person's body as a tool to do/ show them something e.g. using another person's hand to point at a picture.**
- ❖ **Pointing**
- ❖ **Giving another person an object**
- ❖ **Spontaneous imitation – spontaneous imitation of sound or action by the child when no request for imitation has been made**
- ❖ **Screaming**

ities of a child or adult. This means assessing the behaviours in the child's or adult's repertoire that are effective at influencing the behaviour of others and thus finding out if the child or adult can let someone know what they need or want to stop at any given time. One approach to this is very informal and consists simply of keeping a record of the types of behaviour that a child may show that might influence the behaviour of others. This may consist of speech, signs or less formal forms of communication, such as leading someone by the hand, presenting an object, such as a cup, tugging on someone's clothes or types of vocalisations. Box 6.14 gives examples of forms of pragmatic communication that we have observed in children and adults who have Cornelia de Lange Syndrome. Knowing the way in which a child or adult influences the behav-

iour of other people gives us a good starting point for developing the communication system further and thus potentially reducing self-injurious behaviour that seems to be a sort of substitute for other forms of communication.

We have conducted a study of how and whether children with Cornelia de Lange Syndrome try to communicate with their carer when they have no attention or quite demanding forms of attention and the results of this study are shown in Box 6.15. The results of this study showed us that the majority of children with Cornelia de Lange Syndrome have some behaviours that we might think of as communicative and that they do use them at the right times.

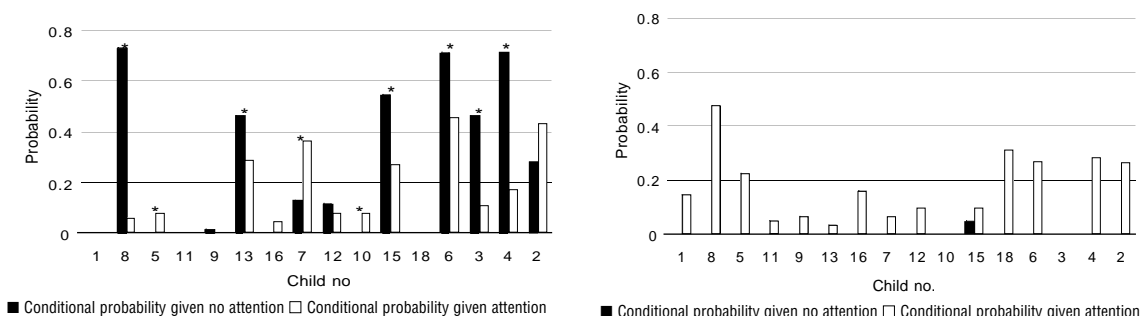
In addition to informally observing and recording the forms of communication, more specific methods can be used. Many of these methods need to be conducted by a Speech and Language Therapist or Clinical Psychologist using standardised tests and questionnaires. Assessment can focus on developmentally related behaviours, such as speech, or more functional aspects of behaviours like those we have described above. Either method will give helpful information but it is important to note that there is a difference between being able to speak or sign and having an effective communication system, that is, one that can effect the behaviours of others.

6.5.2: Adaptive behaviours

There are two important aspects of adaptive behaviour that warrant attention when assessing self-injurious behaviour. The first is an assessment of the individual's ability to control the environment. This means considering the behaviours that the individual has that they may use to satisfy their needs and identifying ways in which any behaviours can be taught such that the individual has an increased range of behaviours and is thus less reliant on others. This area warrants more attention than we are able to give it in this book, consequently the reader is referred to the applied behaviour analysis texts listed in the

Box 6.15: Communication in Cornelia de Lange Syndrome

The two graphs below show the results from a brief analogue study that was carried out with a group of children with Cornelia de Lange Syndrome. In this study conditions of no attention and attention were alternated. The presence of communicative engagement and disengagement behaviours was recorded. The dark bars on the graph represent the probability or likelihood of engagement behaviours occurring during sessions where no attention was given to the individual. The white bars represent the probability or likelihood of engagement behaviours occurring during sessions where attention was given to the individual. A * above the graph represents a statistically significant difference.



Graph 1: Engagement behaviours
 The results indicate that three children (5, 7 & 10) were more likely to display engagement behaviour during attention conditions. Six children (8, 13, 15, 6, 3 & 4) were more likely to show engagement behaviour during the no attention condition. In both cases the presence of engagement behaviour indicates that children with Cornelia de Lange Syndrome have the intent and some ability to communicate with others.

Graph 2: Disengagement behaviours
 All children showed increased levels of disengagement behaviours during attention conditions. Again this shows that the children have the intent and some ability to communicate with others.

Bibliography ^{17, 18}. The most important thing to know is that behavioural or teaching techniques are very powerful in being able to increase the range and use of adaptive behaviours that can lead to greater independence for any individual. The second aspect of assessing adaptive behaviours is less focused on the individual and more focused on the environments in which the individual lives. It may be the case that an individual is heavily reliant on others in order to satisfy their needs not because they do not have the appropriate adaptive behaviours but because they are unable to affect the environment for other reasons. It may be that there are restrictions in the environment that do not allow an individ-

ual to satisfy their needs. So, being unable to access activities, food, drink or other things, although they are available, will mean that the individual is constantly reliant on others to help with access. The restriction in the environment that exists may be present because the environment requires adaptation to cater for an individual's disability or because there are rules that do not allow the individual to control their environment and thus satisfy their needs. When assessing self-injurious behaviour that occurs for psychological reasons, especially social reinforcement, it is critical that the adaptive behavioural repertoire of the individual is considered and that any restrictions on the individual using these

adaptive behaviours to maximum effect are both given attention.

6.5.3: Daily activities and programmes

So far we have focused on the individual's behaviour and the immediate psychological causes for self-injurious behaviour. Before starting any intervention for self-injurious behaviour that is underpinned by reward, it is important to consider the broader context in which an intervention might be conducted. There are three broad areas that warrant attention and form an important backdrop to an intervention. First, a review of the level of occupation and stimulation available to an individual should be considered. This does not just mean a one-off assessment and ensuring that some toys or activities are available. It means ensuring that there is variability in activities that are available and that these are under constant review and that new activities and occupations are constantly available. If an individual has high levels of occupation and stimulation then they are less likely to be reliant on others to mediate their access to reinforcing activities and consequently social reasons for self-injurious behaviour are less likely.

Given the discussion of setting events in Chapter 5 and how they might interact with the presentation of unpleasant tasks, it is important to note when tasks are conducted throughout the day and to evaluate whether these are likely to be associated with setting events that make any task more unpleasant. Good examples of this are presenting tasks when people are tired, during the low period in the early part of the afternoon, or are hungry, prior to meals. Organising the day such that a setting event does not occur alongside an influential antecedent might be important in an intervention and consequently the overall programme needs to be evaluated for these potential interactions.

6.6: Loss of control

In Chapter 5 we considered the possibility that for some individuals who are showing self-injuri-

ous behaviour the psychological or medical reasons that we have outlined so far are less influential and that, for reasons that we do not fully understand, the behaviour seems not to be completely under the control of the individual. We would emphasise that research into self-injurious behaviour that appears not to be under the individual's control has not really been able to explain why this might be, so consequently some of the information presented here is speculative. There is evidence that in Cornelia de Lange Syndrome the self-injury can become severe and it can become difficult to identify any medical and psychological reasons for the behaviour. It is possible that the behaviour has a "compulsive quality" and the individual has reduced control over the behaviour¹⁹.

Assessing self-injurious behaviour to determine whether there is evidence of a loss of control is not easy. We think there are two important factors that might indicate that self-injurious behaviour is not under the control of the individual. The first of these is self-restraint and the second is the presence of some other specific behaviours.

Self-restraint In some individuals who show self-injurious behaviour the person shows a preference for imposed restraint (will clearly prefer to wear items such as splints, gloves or a helmet) or shows self-restraint behaviours that they initiate themselves (such as covering or sitting on their hands). In the first instance a preference for imposed restraint may be seen when individuals actively help in putting on devices such as splints or helmets on and actively seek out these devices if they do not have them on. Additionally, they may become extremely distressed when these devices are removed and appear anxious, tearful or very angry. At the same time, when they do not have the devices they may also try to show forms of self restraint, apparently in an attempt to restrict their own movements.

Self-restraint is different because it does not involve a device that someone else has provided for the individual, and instead the individual has learned a way of controlling their own behav-

Box 6.16: Forms of self-restraint

The table below shows the percentage occurrence of different forms of self-restraint in individuals with Cornelia de Lange syndrome²⁰.

Form of self-restraint	%
Holds onto others/holds onto others clothing	42.6
Holds or squeezes objects	36.2
Wraps self in own clothing or holds onto own clothing	29.8
Chooses to wear particular items of clothing	29.8
Holds hands together, holds onto self	21.3
Positions self to restrain.	14.9
Chooses mechanical restraint	0

This is similar to our results for individuals with intellectual disabilities without Cornelia de Lange syndrome²¹. The table below indicates the percentage occurrence of different forms of self-restraint in this population.

Form of self-restraint	%
Holds or squeezes objects	50%
Holds onto others or others' clothing	41.3
Positions self to restrain	39.1
Wraps self in own clothing	21.7
Holds onto self	21.7
Wears item to discourage SIB	17.4
Chooses mechanical restraint	8.4

our. The forms of self-restraint are numerous and Box 6.16 gives a list of the types of self-restraint that we have observed in individuals with intellectual disability who show self-injurious behaviour and in individuals with Cornelia de Lange Syndrome who show self-injury. Self-restraint can have some similarities to the preference for imposed restraint. Individuals may become distressed if the self-restraint is terminated or if they are showing a form of self-restraint such as covering hands with their sleeves and the clothes that they are offered does not allow this behaviour to occur.

The association with other behaviours. Another indication that self-injurious behaviour may not be completely under the control of the individual or that it may in the future not be under control is the presence of compulsive behaviours. We would emphasise at this point that we do not have strong evidence that this is

the case and that we are speculating that the presence of these compulsive behaviours indicates present or future lack of control. However, our research to date shows that for those individuals with Cornelia de Lange Syndrome who show self-injurious behaviour and compulsive behaviours, self-restraint occurs more than we would expect by chance. It is possible that this is explained by the self-injurious behaviour itself being compulsive, and consequently out of the individual's control, and as a result the individual shows self-restraint in order to limit the behaviour.

6.7: Using assessment information

Once the assessments of these different potential causes of self-injurious behaviour have been conducted it is important to collect the information together and to start to build a model of why

self-injurious behaviour might be occurring. This is called the process of formulation and it is helpful because it will tend to show numerous potential points for intervention. In building a model of self-injurious behaviour it is likely that there will be different levels of strength of evidence for any given cause. It is also quite unlikely that there will be a clear-cut cause that indicates a single specific intervention that needs to be undertaken. It is more likely that there are a number of ways in which an intervention can be put together starting with changes which address the cause for which there is the strongest evidence.

The important thing about the assessment process, and how it is associated to interventions, is being prepared to collect careful information and being prepared to identify a number of causes, prioritise them and address them with interventions one by one whilst carrying out an evaluation of effectiveness. If you feel that a given cause might be influential you should be prepared to experiment to see what happens if you change things in a way that would tell you whether or not something is influential. The second important thing is to keep an open mind in terms of the potential causes of self-injurious behaviour. It is important to note that when we look for the causes of behaviour there is a strong tendency for us to seek information to confirm what we already think. Rather than do this, it is much more important to look at all the evidence and to work systematically through the assessment process from start to finish and to keep looking hard for evidence that contradicts what we think as well as evidence that confirms what we think.

Finally, if you have worked through the assessments and you are unable to find any way in which self-injurious behaviour appears to be linked to any of the reasons that have been described then all is not lost. There are still interventions that we can try and whilst you may not be guided towards a particular intervention you can still work through different interventions keeping a record of their effect on self-injurious behaviour and operate on a trial-and-error basis.

That is, instead of putting into place an intervention because you know the reason for the self-injurious behaviour you can try an intervention and see if it works. There is nothing wrong with this approach when we cannot find causes.

There are two final and important aspects of completing the assessment process. First, you must share the results of the assessment with anyone else who is involved with the child or adult who are showing self-injurious behaviour. You must also be prepared to listen to contradictions from others that indicate that the model you have built of self-injurious behaviour might need to be modified in some way. Finally, whatever model of self-injury you build at one point in time may not be appropriate later on. You must be prepared to change the model over time and to go back and look at specific assessments, repeat assessments and collect more information.

6.8: Summary

Systematic assessment of the causes of self-injurious behaviour underpins successful intervention. The first step is to evaluate whether the self-injury is related to pain and discomfort by looking carefully at the site of the injury, the variability of self-injury across environmental events and the presence of other behaviours that might indicate pain and discomfort. If this reason for self-injury is ruled out, then sensory and social reward should be considered. This will mean collecting informal information, record keeping and observing to examine the association between the self-injury and environmental events. When this process is complete the information should be shared with the key people who have contact with the child ready for the intervention to begin.

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Chapter 7: Interventions for self-injurious behaviour

7.1: Introduction

For any carer or parent of a child or adult who has Cornelia de Lange Syndrome there are two broad issues that relate to interventions for self-injurious behaviour. The first is prevention. Whilst we noted in Chapter 4 that self-injurious behaviour is not necessarily associated with Cornelia de Lange Syndrome any more than we would expect by chance, it is still true that a significant proportion of children and adults with Cornelia de Lange Syndrome show self-injurious behaviour. Therefore, it is important to be aware that self-injury can develop and that proactive prevention of the development of more self-injurious behav-

Box 7.1. Four important things to know about preventing self-injury from developing

- 1. Pain and discomfort can lead to self-injury and the cause needs to be identified quickly and resolved with medical treatment.**
- 2. Find stimulating activities for children to have available so that they do not show mild self-injury to gain stimulation.**
- 3. If you see mild self-injury, such as banging teeth with hands, be aware of how you and others are responding to it. Are you rewarding the behaviour?**
- 4. A functional communication system is important to prevent socially reinforced self-injury developing. Any child or adult must have an effective and reliable way of letting others know what he or she wants to happen next.**

our is important to bear in mind. We hope that a clear understanding of the causes of self-injurious behaviour, outlined in Chapter 5, and the interventions that are described in this chapter should help parents and carers prevent the development of more severe self-injurious behaviour (see Box 7.1). The second issue is interventions for self-injurious behaviour when it is occurring. This is the focus of this chapter in which we describe how to use the assessment information that has been collected to create an intervention that can succeed. Before describing the different interventions that might be implemented on the basis of assessment information, there are some general issues which are important to note.

The first issue is the importance of caring for injuries that result from self-injurious behaviour. It is, of course, important to ensure that any injuries are dealt with appropriately to prevent infection and also to ensure that unseen injury has not taken place. It is particularly important to try to get wounds to heal rapidly to prevent infection but also because the longer a wound is open the more people find it itches and consequently they may respond to this by picking or scratching the wounds and a vicious scratch-itch cycle develops. Covering the wounds and seeking medical advice on promoting healing at this stage is important. The second issue is that it is very important when putting together an intervention that others are involved in this process and all information on the intervention is shared. This means ensuring that all people who come into contact with the person showing self-injury are aware of the intervention strategy that is being used and are constantly updated on any changes that may take place.

It is important to seek professional help and advice at various stages. Some interventions are more easy to implement than others but it is always important to try and work in collaboration with a professional who has experience of implementing interventions. For psychological intervention this kind of professional help should be available from educational psychologists, clinical psychologists and nurses and teachers who may have been trained in behavioural methods (also known as behaviour modification or applied behaviour analysis). There is no doubt that psychological interventions are difficult to implement and a team approach is more likely to lead to success.

It is also important to note that intervention for self-injurious behaviour is more of a process than something that happens once and then does not need to be repeated. We have outlined the process at various points in this book and would emphasise the importance of knowing background information about Cornelia de Lange Syndrome as well as information about self-injurious behaviour in all people who have intellectual disabilities. It is also critical that anyone thinking about an intervention for self-injurious behaviour is thoroughly familiar with the potential causes and has conducted a systematic assessment to try to determine which cause or causes are most applicable to the person they intend to work with.

Once this has been done a plan can be formed and the intervention can be implemented. At this point you are only at the first stage of intervention and it is critical that two things now take place. First, whatever intervention you conduct, you must evaluate the effect of the intervention. This means keeping a regular and systematic record of the self-injury in order to determine whether or not you are being effective. The second is that you persist with an intervention for a reasonable period of time in order to give the intervention time to work. You should not expect rapid and immediate results as it may take time for people to learn different ways of behaving. These two issues are constantly referred to as we

consider the different types of interventions that might be implemented. On the basis of the constant evaluation that is conducted it is extremely important that you are prepared to modify the intervention and try different things. This will be based primarily on your understanding of why the self-injurious behaviour is occurring but will also be based on the results of the evaluation. This overall process of intervention can be repeated many times and it is important that any effective intervention is constantly reviewed and that those involved in the intervention are prepared to change.

In the sections on intervention methods you will see that we mainly target self-injurious behaviour and the way in which this behaviour can be decreased. However, it is important to note that increasing adaptive behaviours (the term 'adaptive behaviour' usually refers to independent living skills and other behaviours such as communication) alongside decreasing self-injurious behaviour is an important aspect of intervention and does seem to lead to longer term change. It is often tempting to focus only on the self-injurious behaviour and not on the adaptive behaviours. We would emphasise that increasing adaptive behaviours is every bit as important as decreasing self-injurious behaviour and we would strongly advise that as much attention is paid to this aspect of the intervention.

In the population of people with intellectual disabilities and, we believe, within Cornelia de Lange Syndrome it is the case that more severe self-injurious behaviour is associated with a greater degree of intellectual disability. Individuals who show self-injurious behaviour are more likely to have poor expressive communication and limited adaptive behaviours. This is important for us to know as it will determine the types of intervention that might be implemented. Because of this association between the greater severity of intellectual disability and self-injurious behaviour, in the section on psychological interventions we tend to focus more on interventions that are appropriate people with severe intellectual disability. However, it is important to

remember that if someone has speech then other types of intervention are available and we include a brief description of these methods.

7.2: Keeping records to evaluate interventions

There is little doubt that the most important aspect of intervening is keeping a record of whether or not the intervention is working. In this section we will describe how these records may be kept and how we can evaluate whether or not our intervention is working.

7.2.1: What to record

It may seem obvious to say that it is the self-injurious behaviour that needs to be recorded but it is important that we define exactly what we are going to record in order that everyone involved records the same thing. There are a number of ways in which we might record self-injurious behaviour. First, we need to define what we mean for any given individual by self-injurious behaviour. It is better to use specific terms and to write out a definition of what we mean so that we can agree when the behaviour is occurring

and when it is not. We call these operational definitions. So, rather than use the term self-injurious behaviour when we are evaluating an intervention we would keep records of, for example, skin-picking or head-banging so that we are clear about the target behaviour. Each of these behaviours might be individually defined so that everyone records the same thing. Box 7.2 gives some examples of operational definitions for self-injurious behaviours.

A second way of recording is that rather than focusing on the behaviour itself we can record or measure the effect of the behaviour. So, we could record the number of scratch marks on someone's arm, the size of an abrasion or whether a new scratch mark has appeared in a given period of time, such as a day. A third way of recording is to make a rating of all the self-injurious behaviour across a given time period. So, we may rate on a scale of 1 (not at all) to 5 (nearly all the time) how frequent the self-injurious behaviour has been in, say, an hour or half a day or a day. (see Box 7.3 for examples of how these records can be kept). These three methods of keeping a record of self-injurious behaviour give us different types of information in order to

Box 7.2: Operational definitions of self-injurious behaviour

The table below provides some examples of operational definitions of different types of self-injurious behaviour that we have used in our research. As you can see they are very specific but this helps make sure we all mean the same thing. It is important to define exactly what we are going to record to ensure that everyone involved agrees on which behaviour has been seen.

Self-injurious behaviour	Operational definition
Body to object banging	Movement of the hand or body down onto an object (excluding body throwing and slapping surface)
Body hitting	Movement of the hand or object down onto the trunk of the body
Body picking	Use of the finger nail(s) to scratch or pick at the body.
Body poking	Pressing the tip of a single finger or thumb into the body.
Eye poking	Pressing the tip of a single finger or thumb into the eye.
Face hitting	Rapid movement of the hand making contact with the face
Face picking	Use of the finger nail(s) to scratch or to pick at the face including cheek and jaw line.
Hand biting	Enclosing and clamping teeth down onto fingers or hand.
Head bang	Movement of head towards and making contact with a surface (e.g. tables, walls floors)

Box 7.3: Keeping records of self-injurious behaviour.

The table below gives an example of how to record the frequency of self-injurious behaviour. The person completing the record indicates the number of times they have seen the behaviour in each hour. A completed record gives us an idea of how frequently self-injury is occurring.

Day	1	2	3	4	5	6	7
Time							
9-10	5	6	4	4	8	7	4
10-11	5	5	4	6	9	5	8
11-12	6	4	4	8	3	6	6
Average	5.3	5	4	6	6.6	6	6

The average is calculated by dividing the total number of self-injurious responses occurring in a day by the number of hours that the person was observed for (e.g. day 2 is $15/3 = 5$; day 3 is $12/3 = 4$). Recording behaviour in this way before and after interventions indicate how successful the intervention has been in reducing the frequency of self-injury.

evaluate an intervention. The methods have different strengths and limitations. Clearly counting the self-injurious behaviour is difficult because we need to carry out almost constant observations (we discuss ways of doing this later in this chapter). However, this is the most accurate way of evaluating an intervention. The other two methods are easier to do but they tend to be less reliable when it comes to evaluation. It is important that you use the most reliable method that you can and that you check that everyone is recording the same thing.

7.2.2: How to record

Just as there are different things that we might record so there are a number of ways in which we can carry out the record keeping for self-injurious behaviour. We can simply count how many times the behaviour occurs within a given time

Box 7.4: Momentary time sampling and partial interval time sampling

Momentary time sampling

Momentary time sampling involves recording occurrences of behaviour and possible antecedents at intervals throughout a day or across several days. The normal procedure is for the observer to have a walkman or watch that beeps every, say, 5th minute to remind them to observe. The observer then looks at the target individual for a few seconds and records a) whether or not self-injury (or other problem behaviour) is occurring and b) what else is happening i.e. attention, demand, playing alone etc. The observer then looks away and does not observe the individual until the next bleep. The beeps can be spaced as far apart as is practical, although the ideal interval size will depend on the frequency of the behaviour being observed. This is a useful technique as it ensures that observations are random and not just taking place because the target behaviour is occurring. It is also easy to carry out and does not require lengthy observations of several hours at a time.

Partial interval sampling

Partial interval sampling works in a similar way to momentary sampling but in this case, observations are continuous across the whole period. The session is split into sections (ranging from 10 seconds to several minutes, depending on the behaviour) and as before, a walkman or watch is used to signal moving from one section to the next. Target behaviours and environmental events are ticked off if they occur at any point during a particular section. This process is more time consuming than momentary time sampling but can be more accurate if the time periods for momentary time sampling are too long.

period, for example, an hour, a day, or a week. This is called a frequency count and the record tends to be accurate. However, the method involves constant observation and this may not

always be practical and we discuss methods of what is known as time sampling below. Whilst frequency counts are the most common way of recording they may not give the most accurate picture of behaviours that have a very low frequency but tend to occur for long periods of time. An example this is eye pressing which may only occur once or twice in an hour but when it does occur it could occur for 20 minute periods. Generally speaking it is better to record how long these sorts of behaviours occur for, as we really want to decrease the amount of time spent in the behaviour. Frequency counts are more useful for behaviours that have a short and stable duration.

It is of course often impractical to observe someone all the time and consequently we can use methods of sampling in order to keep an accurate record. There are two ways in which we can sample. One is that instead of watching someone for a whole day we could watch them for, say, three half-hour periods throughout a day in order to record the level of self-injurious behaviour. A second way in which we can sample is that when we are observing we can use some time sampling methods in order to get accurate records. There are two methods of time sampling that we would recommend: momentary time sampling and partial interval time sampling and these are described in box 7.4. In both of these methods we would use a personal cassette player with a pre-recorded prompt on the tape to make a record of the self-injurious behaviour on paper. This will give us a reasonably accurate record of the self-injury without having to observe the person all the time.

7.2.3: Using the data that you collect

The best way to evaluate the effects of an intervention is to keep a record of the self-injurious behaviour before the intervention starts (this is called the baseline) and then to keep the same record whilst the intervention is running. The most useful thing to do is to plot the information on a graph and then you have a good visual picture of whether the behaviour is increasing, stay-

ing at the same level or decreasing. It is also useful to use the graph to keep a record of any unforeseen incidents that you think might have affected the self-injurious behaviour so that you can see whether or not a pattern emerges. Box 7.5 describes how to plot a graph based on the information you have collected.

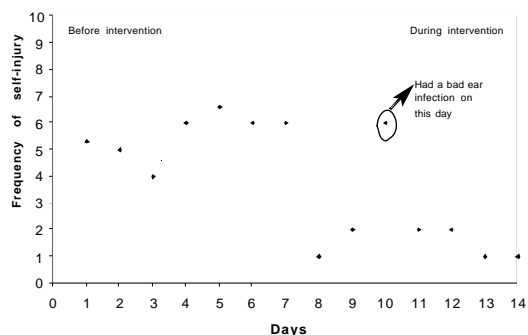
7.3: Interventions for self-injury related to medical causes

In Chapter 2 we described the medical conditions that are more commonly associated with Cornelia de Lange Syndrome and that may give rise to pain and discomfort. It should be noted that whilst these medical conditions are associated with Cornelia de Lange Syndrome it is, of

Box 7.5: Plotting graphs

Box 7.2 gave an example of how to record the frequency of self-injurious behaviour. Sometimes it is much easier to see patterns in the data when you plot it in a graph. In particular this is useful for seeing how successful an intervention has been at reducing the severity of self-injury in a particular individual.

The graph below is an example of how you might plot this data. It shows the frequency of self-injury before and after the implementation of an intervention.



The graph enables us to see that self-injury is markedly decreased after the intervention apart from on day 10 when it increases again. As is indicated on the graph the individual had a bad ear infection on this day which might explain this transient increase in behaviour.

course, likely that children and adults with the syndrome will, at some time have medical conditions that affect anybody. In Chapter 5 we discussed how we think self-injurious behaviour is related to pain and discomfort and in Chapter 6 we discussed how a potential relationship between a medical condition and self-injurious behaviour can be assessed. If it is thought that pain and discomfort might be related to self-injurious behaviour from the results of an assessment then the most effective way of trying to decrease the self-injury is to deal with the medical cause. This means involving medical staff who are prepared to systematically evaluate potential sources of pain and discomfort and try methods to relieve the resultant pain and discomfort if the root cause cannot be cured. Whilst this strategy seems obvious we believe it is very important that it is vigorously pursued and that all possible medical causes are ruled out before implementing psychological interventions. The Cornelia de Lange Syndrome website is a good resource for learning more about the treatment protocols for medical conditions that are associated with Cornelia de Lange Syndrome.

We also noted in Chapter 2 the possibility that individuals with Cornelia de Lange Syndrome might have a peripheral sensory neuropathy. This may give rise to unusual sensations and feelings in the feet, legs, hands, fingers and arms and this may lead to biting or scratching of the area where this sensation is experienced. From the research literature that we have examined there is very little indication that there is an effective intervention for these sensations. However, there is a case report in a medical journal that describes hand and arm massage as effective in reducing self-injurious behaviour in a woman who has Cornelia de Lange Syndrome¹ (see Box 7.6). It is possible that this massage worked by in some way alleviating or cancelling out these sensations that were experienced by the individual. This seems a possible intervention and at present the only way to know whether it may work is to systematically evaluate the effect of this intervention in the way we have described above.

Box 7.6: Massage for very severe self-injurious behaviour in a girl with Cornelia de Lange Syndrome

This case report was presented by Dossetor, Couryer & Nicol in 1991¹. A young girl (LH) with Cornelia de Lange syndrome was described who, at age 14, had been displaying severe self-injury for ten years. Medication had been tried but had been unsuccessful in treating the behaviour. Only splints and a helmet could prevent LH from engaging in self-injurious behaviour. Functional analysis had failed to reveal any consequences of the behaviour. As a part of a new treatment programme LH received a 30 minute massage twice a day. From the first day, she enjoyed and relaxed with the massage. It had a beneficial effect on her behaviour and mood for the rest of the day. After the third day LH indicated that she wanted to massage others as well. Thus a form of reciprocal play developed. After six months, her self-injurious behaviour improved so much that she was taken off all medication for the first time in ten years. She wore no splints or helmet and her injuries healed. The authors indicated that the improvements had been maintained 18 months later. Any relapses that had occurred had been mild.

7.4: Interventions for self-injury related to sensory reinforcement

In Chapter 5 we noted that there is good evidence that self-injurious behaviour can occur simply because the sensory or perceptual effects of self-injury are in some way rewarding. The type of reward can be of two types. First, there can be alleviation of pain and discomfort (this process would be referred to as sensory negative reinforcement) and interventions related to medical causes can be explained by this process. Second there can be reward that is positive from the stimulation that comes from the self-injury. This seems most likely for self-injurious behaviour that involves mild eye pressing and perhaps finger

Box 7.7 Providing stimulation in a more adaptive way

Judith Favell and colleagues described how Dane, a 14 year old boy with profound intellectual disability with light\dark perception only, showed eye poking for approximately 40% of the time³. When Dane was provided with toys with visually striking properties e.g. brightly coloured toys, mirrors and translucent coloured shapes the eye poking decreased to about 10% of the time. One interpretation of this decrease is that as visual stimulation comes from the toys there is no longer a need for eye poking.

picking and hand biting. Although it should be noted that there is no evidence that the type of self-injurious behaviour is necessarily related to any particular cause, and any form of self-injury can be reinforced by sensory consequences.

For self-injury that appears to be related to the positive sensory consequences there are three basic strategies. These involve increasing alternative forms of sensory input, decreasing the sensory input that arises from the behaviour and increasing the available rewards for not showing the behaviour. Before considering any of these interventions it is important to think through a number of issues. First, it has been argued that some mildly self-injurious behaviours are developmentally appropriate². This means that all children at some time will suck and bite their hands and if you watch any child then at some point you may see them scratch or, for example, bang their head. For some of these behaviours, particularly mouthing hands, this is a way in which the very young child will explore their environment. However, the problem is that because development may be much slower and may plateau at an earlier stage for children with Cornelia de Lange Syndrome, then these behaviours may stay in the child's repertoire for a much longer period.

The second issue is deciding whether or not to intervene. For mild self-injurious behaviours that are not causing considerable tissue damage it

is important to think through what the person will do if they are not showing these behaviours. In other words is there a good alternative to the behaviour that you may want to decrease. For behaviours that are injurious there is clearly a good case to be made for trying to decrease the behaviour. However, for behaviours that are extremely mild and no alternative can be developed then there is perhaps a less strong case. The other factors to take into account when considering whether to intervene is the extent to which the behaviour is additionally handicapping for an individual. Some behaviours can make children look very unusual and for this reason it may be important to try to decrease the behaviour.

7.4.1: Increasing a specific type of sensory reinforcement

When self-injurious behaviour is shown because the sensory reinforcement that is experienced is positively rewarding, then increasing the type of sensory reinforcement can lead to decreases in self-injurious behaviour. In practice this means trying to understand the kind of sensation or perception that the child gets from the self-injurious behaviour and then trying to present this kind of stimulation in a different way. (See Box 7.7) This is, of course, not always practical or easy to do, for example for head-banging. However, for hand biting or hand mouthing then providing different activities that stimulate the child's hands and mouth can be effective. The real task is finding an activity that the child likes as much, and preferably more, than the effect of the self-injurious behaviour whilst ensuring that this does not look as unusual as the behaviour and that the child does not tire of the activity quickly. This is where a process of trial and error and an active imagination is important.

7.4.2: Increasing the general level of sensory reinforcement

The second strategy is to try to increase the amount and variety of sensory reinforcement that

is available for any child or adult as much as is possible and practical. There are a number of ways in which this can be achieved. One way is by ensuring that there is a high turnover of sensory activities. This means that the child's activities are changed at very regular intervals such that the child does not become bored with the activity that is available to them and reverts to self-injurious behaviour to seek stimulation. The second strategy is to ensure that there are a variety of sensory activities available to the child. This means that rather than providing activities or toys that are all stimulating to the child in the same way, that different toys and activities giving very different effects are made available. The third strategy is to try to increase the overall level of activity that the child is experiencing. This means ensuring that the level of stimulation is generally kept high so that the child does not need to revert to self-injurious behaviour to provide stimulation. Inevitably this means a good deal of trial-and-error to find activities that child finds reinforcing. Low input, high return toys are particularly good for this purpose and local toy libraries for children with intellectual disabilities can be a very good resource to find activities and try them out (see Box 7.8).

Box 7.8: Toys that can provide stimulation to compete with behaviour

Glyn Murphy and her colleagues described how David, a 14 year old boy with profound disabilities, partial sight and hearing problems, showed high levels of a self-stimulatory behaviour (rocking) even when he had toys to play with⁴. However, when one of the toys was adapted to vibrate when it was touched, the self-stimulatory behaviour went down from 85% of the time to 15% of the time and active toy touching increased from 6% of the time to 94% of the time. Although the rocking is not self-injurious, this example does show how providing stimulating toys can effectively compete with the stimulation that comes from a repetitive behaviour. A similar finding was reported by Jon Bailey and Lee Meyerson in 1970 for the effect of vibration on headbanging.

Box 7.9: Changing the consequences for self-stimulatory behaviour

Arnold Rincover first reported the method of sensory extinction in 1978. He described Reggie, 14, who had profound intellectual disability and visual impairments who would spin objects, particularly plates (!) on hard surfaces in a repetitive way⁵. When the sound from the plate spinning was muffled, by padding the table top, the amount of time Reggie engaged in plate spinning dropped from about 60% of the time to nearly zero. Rincover argued that this was because the behaviour was rewarded by the noise of the plate spinning and when this was muffled the behaviour was extinguished.

7.4.3: Sensory extinction

Sensory extinction was first described in the 1970's and referred to a process whereby the sensory consequences of a behaviour are eradicated such that the reward no longer occurs and consequently the behaviour will decrease. This has been demonstrated for both self-stimulatory behaviours, such as plate spinning and hand-waving, and some self-injurious behaviours (see Box 7.9). However, it should be noted that the evidence for self-injurious behaviour is rather limited. Basically, the procedure consists of removing any sensory consequences to the self-injurious act. In practice this may mean padding or covering the area that is targeted for self-injury such that the individual does not experience any feedback whatsoever. There are two major problems with this technique. The first is that the person may well try to remove the pad or protection in order to seek the stimulation and the second is that the padding often needs to be thick and obtrusive and this may make the child look unusual. We would certainly recommend that if this type of intervention is tried then it is important to combine the intervention with the two strategies that have been described above (increasing the specific type of sensory rein-

Box 7.10: Four important things to know about intervening for self-injury maintained by sensory stimulation

- 1. Providing an alternative form of stimulation can decrease self-injury, the closer the type of stimulation is to the effect of self-injury the better.**
- 2. Alternative forms of stimulation, such as toys, should be low input with high return, changed regularly and with new forms introduced often.**
- 3. Present rewards when the person touches toys or other stimulating objects. These can be from you but might be in the object itself (objects that vibrate or light up when touched).**
- 4. Present varied and stimulating activities throughout a day.**

forcement and increasing the general level of sensory reinforcement) and/or the reinforcement competition strategy that we describe next.

The other way in which the sensory consequences of self-injury can be minimised is by a carer preventing the responses when they occur (so called responses prevention) There are a number of problems with this method. First, for high rate behaviours this can be very demanding of carers. Second, there is a risk that by providing a social response to the behaviour it may become socially reinforced, although this can be minimised by the social response being very cool (no speech, no eye contact). Third, although the child may learn not to show a behaviour he or she will not necessarily learn what to do instead. For these reasons this type of intervention needs to be implemented with caution and certainly alongside another strategy.

7.4.4: Reinforcement competition

This strategy can be used in addition to the interventions described above and is considered in much more detail below. Basically, the interven-

tion consists of presenting a reward for the individual when they are either not showing the self-injurious behaviour or they have not shown the behaviour for a given period of time. The different ways in which this can be achieved are described in Section 7.8.2 in which the importance of identifying the right reward and frequent changes of rewards is also described.

For a summary of the main points about self-injury that is rewarded by sensory stimulation see Box 7.10.

7.5: Interventions for self-injury related to social reinforcement

In Chapter 5 we focused on the fact that self-injury can occur because of the social reinforcement that is presented by others. There is a good deal of evidence in the research literature that this is a common cause of self-injurious behaviour in people with intellectual disabilities and from our research we have no evidence to suggest that the cause is not appropriate for people with Cornelia de Lange Syndrome (see Boxes 6.2 and 6.12). In the broader research literature on social reinforcement in people with intellectual disabilities the evidence is that for approximately 70 percent of people the cause of self-injurious behaviour is likely to be social reinforcement (see Box 7.11). Additionally, there is good evi-

Box 7.11: How common is social reinforcement for self-injurious behaviour

Brian Iwata and colleagues carried out a review of all the studies of self-injury that had used analogue methods as an assessment for whether the self-injury was socially reinforced⁶. 152 children and adults had taken part in the assessments and the results showed that 23% showed self-injury that was rewarded by attention, 35% by escape from tasks and just under 10% for other social reasons. 20% were thought to show self-injury because of the sensory stimulation but the evidence for this figure is less strong.

dence that when the cause is social reinforcement psychological interventions can be effective, although they can be rather difficult to sustain over long periods of time. In this section we will describe interventions for self-injury that is related to social reinforcement and, just as for the section on assessment, we will differentiate between self-injury that occurs because of a positive social reinforcement process (i.e. self-injury that tends to be rewarded by social attention or by tangible items such as drinks or activities) and self-injury that occurs because of negative reinforcement (i.e. self-injury that tends to be rewarded by escape from task demands or social contact).

7.5.1: Self-injury maintained by positive social reinforcement

There are two general strategies that can be implemented when the results of an assessment show that self-injurious behaviour is maintained by the social attention that follows the self-injury or the presentation of more tangible things such as food, drinks or activities. The first is to try to generally increase access to these reinforcers. That is, to increase the overall level of attention from others and increase the availability of the more tangible items that have been reinforcing the self-injurious behaviour. There are a number of specific ways in which this can be done and these are described below. The second general strategy attends to the issue that attention is extremely rewarding to almost any child or adult and that it cannot always be presented. It is important to ask the question why attention is so rewarding and reinforcing and consequently understand why the person wants attention at such regular intervals.

Apart from the intrinsically rewarding nature of social attention, it is likely that other people often act as the link between the individual who shows self-injurious behaviour and things that they wish to access. That is, people with severe intellectual disabilities will inevitably be highly reliant on other people to satisfy their needs. The

general strategy therefore is one of trying to increase levels of adaptive behaviour and remove the barriers that might exist in the environment that limit an individual's access to the things that they want or need and thus require other people to present. Whilst this may not seem to be a priority when self-injurious behaviour is occurring, it is an important background strategy that will help any psychological intervention to be effective. We fully acknowledge that increasing adaptive behaviour is difficult when working with people with severe or profound intellectual disability. However, there is good evidence that people can acquire new skills when the precision teaching methods of applied behaviour analysis are employed with consistency over time. We should not make the assumption that any child, no matter how disabled, cannot learn new skills. This is particularly important with regard to communication and we discuss this in more detail below.

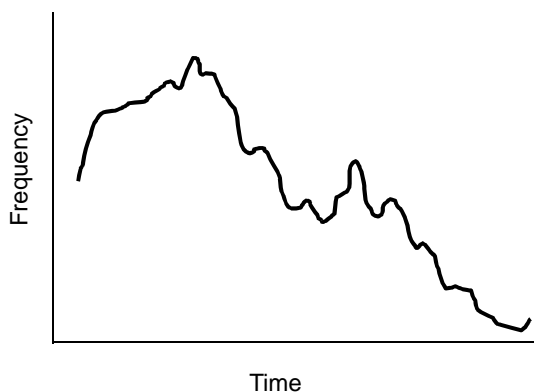
7.5.1.1: Changing the consequences

If the results of an assessment show that the self-injurious behaviour is caused and maintained by the attention that follows the self-injury then it is important to change this consequence (see Box 7.12). There is very strong evidence in the research literature that doing so can decrease self-injurious behaviour⁷. However, this strategy

Box 7.12: The importance of what happens after self-injury

It is very important to find out what happens after self-injury and to think about whether it might be a reward. The most common rewards are social attention and escape from tasks and these can take many forms. Telling someone off, even by shouting, can be rewarding, as can giving eye contact or trying to distract someone from the behaviour by presenting alternative activities. It is critical that you check whether things that happen after self-injury might be rewarding. If there is a chance that it is, then the response must be changed.

Box 7.13: Extinction burst: Things get worse before they get better.



The graph above shows what happened over a 45 minute period when we did not present the normally occurring social reward for a problem behaviour that was being shown by a 14 year old girl (in this example this was aggression)⁹. Within about 10 minutes the behaviour increased in frequency by about 50% and the aggression was also more intense (harder hits and hair pulls). It took about 45 minutes before the extinction burst was over.

should never be used alone and there are a number of issues that need to be carefully thought through before trying to change the consequences. When self-injurious behaviour is caused and maintained by the attention that occurs after the self-injury the temptation is to stop presenting this attention because this should lead to decreases in the self-injurious behaviour. Whilst it is certainly true that the self-injurious behaviour will eventually decrease there are two basic problems with this strategy.

The first is known as an extinction burst⁸. That is when a behaviour has been reinforced in the past and the reinforcement is withdrawn the behaviour does not stop immediately. Rather it increases in both frequency and intensity before finally stopping. Box 7.13 shows an example of an extinction burst. The problem of an extinction burst is evident in our daily lives. If you think

about the last time you lost something you will almost certainly have repeatedly returned to the place where the thing was even though you now know it is not there. It is very similar for a child or adult who has been used to receiving attention after showing self-injurious behaviour. Even though in the short term the reward does not come they will continue to show the behaviour, often more vigorously, for some time before stopping. The problem here of course is that a good deal of injury can be experienced by the child during this extinction burst. This procedure of extinction therefore needs careful consideration before being implemented. It is possible to use protective devices when trying this strategy in order to protect the person from any injury. We would strongly recommend that professional help is sought before considering that this kind of intervention.

The second problem with simply stopping a reward, such as attention, that usually follows the self-injurious behaviour is that the child or adult no longer has any way of gaining the reward. So, whilst self-injury has stopped, the need that the child or adult has for attention has not been removed and then two things may happen. The first is that a different behaviour, most likely a problem behaviour if the person has a limited repertoire of adaptive behaviours, may replace the self-injurious behaviour and the second is that the self-injury may simply come back at a later date. Therefore, when thinking about the use of extinction it is critical that the procedure is combined with a way in which alternative behaviours that can replace the self-injury are taught.

7.5.1.2: Increasing alternative behaviours

In the section on changing the consequences we clearly identified a problem with the strategy of simply stopping the reinforcement that usually follows self-injurious behaviour. If we do this we leave the person without a way of gaining the thing that they want or need. In the research literature there is very strong evidence that teaching a form of communication that can gain attention

Box 7.14: Five important things to know about intervening for self-injury maintained by attention.

- 1. Modify your response to the self-injury. If you must respond, to protect and restrict, be 'cool', no speech, no eye contact, no fun!**
- 2. Present positive and fun attention when self-injury is not occurring. Set a timer to remind you.**
- 3. Find or teach a communicative response that can get attention from you and others. Make sure everyone responds to it.**
- 4. Increase independence to give control to the person and reduce their reliance on your attention.**
- 5. Make sure everyone is doing the same thing.**

under these circumstances can be very effective and it is critical that if the function of self-injurious behaviour is to gain attention then the child has a reliable and effective way of communicating that they want attention. We cannot emphasise enough the importance of trying to increase the child's ability to effectively influence the behaviour of others under these circumstances.

Below we consider the various forms of communication that can be taught to a child in order to help to decrease a behaviour that occurs because it is positively reinforced. Whilst this approach focuses on the child learning to communicate their need for attention to others, it is important to remember that self-injurious behaviour can be positively rewarded by things other than attention. So, it is important that the child is able to communicate a need for more tangible items, such as drinks, food or activities, or that they are able to access them in some other way. There is some research evidence that children with severe intellectual disabilities can learn to use micro-switches that can control aspects of their immediate environment such that they are able to satisfy their needs without an adult being present (see Box 7.15). Whilst this research is at an early stage there is no reason why children

and adults with Cornelia de Lange Syndrome should not also be able to learn in precisely the same way. The more control the child or adult has over their environment and the better their ability to control and acquire the things that they want and need the less they will need to rely on self-injurious behaviour.

For a summary of the main points about self-injury that is rewarded by social attention see Box 7.14.

7.5.2: Self-injury maintained by negative reinforcement

We described in Chapter 5 how self-injurious behaviour may occur when an aversive event or task is presented to the individual and that the self-injury is then reinforced or rewarded by the removal of the aversive event or task. Under these circumstances there are a number of strategies that can be used to try to manage the behaviour and decrease the self-injury in the long term.

7.5.2.1: Reducing the aversive nature of tasks and demands

First it is important to think through why any task may be experienced as aversive or unpleasant. There can be a number of reasons for this and trying to see the task from the child or adult's point of view is an important way of trying to

Box 7.15: Taking control of the environment

In 1989 Mark Steege and his colleagues described Ron, an 8 year old boy who was non-verbal and nonambulant, who self-injured by biting his hand and it appeared that he did so in order to gain some form of stimulation¹⁰. Ron was taught to activate a microswitch so that he could control what was happening in his environment. In fact the switch would activate a radio and a fan. This led to a decrease in his self-injury from about 50% of the time to near zero levels. The nice part of this intervention is that Ron could choose what he wanted and he did not have to rely on others.

Box 7.16: A graded approach to a 'feared' stimuli

A colleague of ours described an intervention for a young boy who showed self-injury mainly when he was being taken toward the toilet. In the past the toilet had been associated with painful attempts to pass faeces, due to constipation, and although this problem was now resolved his avoidance of the toilet persisted and he escaped by self-injuring. The intervention consisted of first rewarding the boy for walking a short distance toward the toilet (but going no further, turning around and going away) and then for getting a bit closer, then touching the door, then opening the door and so on. It took time but eventually the boy was again able to use the toilet and did not self-injure when being taken.

understand why escape from the task is a big reward. Often the tasks that might be presented to children and adults with a severe intellectual disability are not necessarily intrinsically rewarding or the rewards are so long term that the activity is not experienced as worthwhile. It is always worth considering any task or event and asking yourself what is in it for the child?

Sometimes an event is experienced as aversive simply because it evokes fear or anxiety. We may not know how this came about in the first place but often it is because a particular event was associated with a very unpleasant experience at some time in the past and consequently the event now evokes fear. When the child is exposed to this aversive event they may experience the fear or anxiety that occurred in the past and show self-injurious behaviour until they are removed from the situation or the situation is removed from them.

Under these circumstances we can draw on the methods that are normally used to treat phobias in order to try to reduce the degree to which the child experiences the situation as fear or anxiety provoking. One strategy that may be used is called graded exposure. This means closely examining a feared event or situation and break-

ing it down into small components. Then the person can be gradually exposed to small parts of the event or situation and rewarded for not showing the behaviour when they are in this situation. An example of this kind of intervention is shown in Box 7.16. A second strategy that can be used for fear and anxiety provoking events is called "flooding". This refers to exposing the person to the situation and not allowing them to remove themselves from the situation until the fear and anxiety have decreased. This is a very effective intervention for phobias. However, it is likely that if this intervention is tried then the self-injurious behaviour might increase dramatically, because of the extinction burst that was described above and in Box 7.13, in the short term and consequently this is a difficult strategy to employ.

The research literature shows that for negatively reinforced behaviour the type of event that will lead to self-injurious behaviour in children and adults with intellectual disabilities is an unpleasant or unwanted task or demand¹¹. Under these circumstances we may well expect to see more self-injurious behaviour in teaching settings and in one-to-one and school environments. Again, it is important to think through why the child might find the tasks aversive or unpleasant. If we can do this then we may be able to make the task a more positive experience and consequently the child will not show self-injurious behaviour to escape. There are a number of ways in which we can look at tasks and think about why they are aversive.

First, tasks may be experienced as too difficult and need breaking down into small steps in order that there are more opportunities to reinforce the child. Part of this process is called a task analysis and is a common method that is used to help in teaching adaptive behaviours for children and adults with severe intellectual disabilities. An example of a task analysis is given Box 7.17. Once the task analysis has been completed then it is possible to reward the child for completing each component of a task rather than only at the end of the long task. Over time it is

possible to put components of the task together and consequently only present the reward, for example, when two or three components have been completed. This process of reward fading helps to move towards a more natural process of teaching where the reward comes when a task is complete.

It is also important to consider other aspects of the task that may make the event aversive and consequently lead to a burst of self-injurious behaviour. There is some evidence that a very high rate of demands as opposed to few demands that are spaced by a good period of time is experienced as more aversive and thus more likely to lead to self-injurious behaviour¹¹. Similarly, the kinds of prompts that are given to a child or adult to complete a task may also be experienced as more or less aversive. Physical prompts that are high rate and firm may well be experienced as much more unpleasant than more gentle prompts. A trial-and-error process of the components of any teaching session should be able to reveal those parts of task demands that

Box 7.17: Breaking a task down into small steps

Here is an example of how hand washing can be broken down into small steps so that, initially, each one can be rewarded. Later rewards could be given every two steps, then three and so on. This is called forward chaining. A different method is backward chaining. Here everything would be done for the person except the last step. The person is prompted to do the last step and then rewarded. Next time the person is prompted to do the last two steps and then rewarded, then the last three and so on. The advantage of this method is that the reward naturally comes at the end of the task.

1. Turn on tap. 2. Wet hands. 3. Turn off tap. 4. Pick up soap. 5. Rub soap on hands. 6. Put soap back. 7. Turn on tap. 8. Rinse hands. 9. Turn off tap. 10. Pick up towel. 11. Dry hands. 12 Return towel (or, if you are male, drop on the floor for someone else to pick up).

Box 7.18: A little bit longer each time

When children or adults will not take part in any teaching activity, even for a short time, then one strategy to use is “shaping” in which the person is rewarded for spending increasing longer amounts of time taking part in a task. So, first of all you might only reward someone for sitting down at the table for a second or two, then when they have sat for, say, 20 seconds, then a minute, then two and so on. You can also use the same method for increasing the amount of demands. You could present a reward for just touching the task for a second, then for picking up part of the task, then completion of a small bit of the task and so on. You may need to use some prompts initially but these can be faded away to verbal instructions.

are experienced as the most aversive and can be modified in some way.

There are some general aspects of teaching sessions or task demands that can make the experience more pleasant and thus less likely to evoke self-injurious behaviour when it is reinforced by escape from a teaching situation. First, the amount of reward available for participating in small aspects of the teaching session needs to be high and reliably presented. The second is that when resistance to teaching sessions is prominent it is possible to gradually increase the level of demand or the time for which the session is conducted gradually when combined with high levels of reward and reinforcement. (See Box 7.18) Gradual increase and patience is the key. Finally, there is a growing body of research that shows that locating more difficult tasks and events within a stream of easier tasks leads to lower rates of problem behaviour and this includes self-injurious behaviour¹².

So, there are a number of ways in which it is possible to decrease the aversiveness of a task or event such that when self-injurious behaviour is reinforced by escape, the behaviour is made less likely to occur. All of these strategies need considering in the broader context of the importance

of increasing adaptive behaviour in all people with learning disabilities and, for children, the school curriculum and setting. Additionally, it is important to consider whether some tasks or aversive events are really necessary and whether there are alternative solutions to the person acquiring a particular skill when they find the teaching of that skill or adaptive behaviour highly unpleasant.

In addition to these specific strategies that might be adopted to make tasks less aversive there are two general things that might be considered and for which there is research evidence that they are helpful. The first is the importance of task variety. Everyone has the experience that repetitive tasks with very little variability are more boring and consequently more aversive than a series of different tasks. Second, there is growing evidence that allowing people choice over the type of tasks that they will undertake does lead to less problem behaviour than usually occurs in response to demands. Whilst these general strategies seem obvious, it is well worth keeping an eye on whether there are varied tasks being presented and whether the individual has choice over what kind of task they are carrying out.

7.5.2.2: Changing the consequences

As we noted in the section on changing the consequences for self-injurious behaviour that is rewarded by attention or tangible events, there can be problems with simply not presenting a reward. The situation is no different for self-injurious behaviour that occurs when there are aversive tasks or events and the reward is escape from these. There is certainly evidence that when self-injurious behaviour occurs during an aversive task or event, and thus the function is escape from the event, that not allowing the person to escape i.e. continuing to present the task or event will, eventually lead to a decrease in a self-injurious behaviour. This procedure is called escape extinction and the same problems that we have previously described above will occur. That

Box 7.19: Five important things to know about decreasing self-injury that occurs when tasks are presented

- 1. Try not to remove the task when self-injury occurs. If you must, to protect and restrict, come back to the task for a brief time and stop when there has been no self-injury.**
- 2. Break the task into a series of smaller tasks, give big rewards for completion or tries, give time between prompts and check the prompts are not unpleasant.**
- 3. Find or teach a communicative response that can tell you the person wants the task to stop and respond to this.**
- 4. If the communicative response for 'stop' happens too frequently (!) set a timer with the person and only respond after the timer gone off. Gradually increase the time.**
- 5. Make sure everyone is doing the same thing.**

is the behaviour will increase dramatically in both frequency and intensity prior to the eventual decrease of self-injury. Once again if this kind of intervention is considered then it is important to think through the safety issues and the likelihood that it is possible to persist with the task demands with significant injury. It is important to seek advice when considering this kind of intervention and it is critical that the intervention is considered in conjunction with those described above in which the nature of the task demand is modified and also with an increase in alternative behaviours being programmed.

For a summary of the main points about self-injury that occurs when tasks are presented see Box 7.19.

7.5.2.3: Increasing alternative behaviours

We noted above that when behaviour is reinforced by others then simply removing that the behaviour from a child or adult's repertoire will mean that they will then be unable to satisfy their personal needs. The situation is no different for

behaviour that is reinforced by escape from aversive events or tasks. Under these circumstances it is equally important that the ways in which a child can express the need for a task to stop or be removed is attended to. Inevitably, this means trying to increase the ability of the child to communicate that his or her experience in that a task as aversive. In Section 7.6 we consider ways in which an appropriate form of communication can be increased.

7.5.2.4: Interactions with setting events

We have previously discussed the way in which what we referred to as “setting events” might interact with demands. Put more simply this means that when we make demands on anybody, the demands will be experienced as more aversive, and thus more likely to lead to self-injurious behaviour, if there is a setting event present such as fatigue, low mood, pain or discomfort. (See Box 7.20). The identification of these setting events is helpful for intervention planning and trying to identify behavioral predictors of setting events such as low mood, pain and discomfort and fatigue are important. The reason for this is that when these setting events are evident then clearly it is not a good time to introduce aversive tasks. If these aversive tasks are introduced at this time they

will be much more likely to evoke self-injurious behaviour if it is reinforced by escape from these tasks. It is important therefore to have programmes of activities that are flexible and will allow for the opportunity for the child to participate in less aversive tasks if the setting events are evident.

In the longer term it is important to try to modify and eliminate these setting events such that they are not present and thus cannot interact with aversive task demands. Attending to sleep difficulties that may give rise to fatigue, pain and discomfort are good examples of how general interventions may help with self-injurious behaviour. There are a number of methods for dealing with sleep difficulties and information on these can be found in some of the books in the Bibliography. Finally, we would note that the periods after a meal times are perhaps a special case for some children and adults who have Cornelia de Lange Syndrome and are experiencing gastrointestinal reflux. It is certainly true that people who are experiencing reflux do experience pain and discomfort in the oesophagus, upper chest and throat. At these times people may find the presentation of any tasks or other aversive events as even more unpleasant than usual and consequently this might lead to self-injurious behaviour.

Box 7.20: An interaction between a task demand and a setting event

Mark O'Reilly and colleagues describe how a child with Williams syndrome would show problem behaviour when there was background noise whilst tasks were being presented to him but there was no problem behaviour when there was no background noise with tasks or background noise but no tasks¹³. When the child was given ear plugs the problem behaviour decreased. This is interesting because it shows how a feature of a syndrome (hyperacusis, or sensitivity to noise, is a feature of Williams syndrome) interacts with an environmental event, tasks, to increase problem behaviour.

7.6: Functional communication training

Since the mid-1980s there have been repeated demonstrations in the research literature that improving the functional communication of children and adults with learning disabilities can lead to a decrease in different forms of challenging behaviour¹⁴. However, the research literature is also a very clear about two things. First, functional communication training is only really effective when the assessment of self-injurious behaviour shows that the self-injury is reinforced either by social positive reinforcement of attention or tangible items or when it is reinforced by social negative reinforcement of escape from task demands or other types of social interaction. Second, that it is extremely important that

the communication that is taught is matched to the reason that the self-injurious behaviour occurs. In effect this means that when the assessment shows that self-injurious behaviour is maintained by attention or access to more tangible items then the communication must be able to do exactly the same thing. Similarly if the self-injurious behaviour is maintained by escape from task demands or other aversive events then again, the functional communication must have the same effect. (See Box 7.21). These two principles have been repeatedly demonstrated and consequently we would again emphasise the importance of the assessment process that we outlined in Chapter 6.

7.6.1: Precursor behaviours and Functional Communication Training

Before describing the various forms of functional communication that can be taught and some of the principles that should be considered when teaching Functional Communication Training there is some recent research evidence that we think might prove to be important. It is important in Functional Communication Training to pick the right time to prompt the person to show the functionally communicative behaviour. Obviously, we want to do this at precisely the time that they would usually self-injure in order to gain the reinforcement thus displacing the self-injury. However, also we do not want to present the reinforcement for the functional communication when the self-injury has occurred as this may reinforce this behaviour. Recently it has been demonstrated that self-injurious behaviour does not always occur in isolation¹⁶. Rather, there are some behaviours that will happen just before self-injury, especially when it is reinforced by social processes. This means that just prior to self-injury occurring there are what are called “precursor behaviours” that may often be observed. These behaviours may take the form of attempts to attract someone’s attention, for example, vocalizations or increased signs of agitation or mild forms of self-injury prior to the

Box 7.21: Saying the right thing at the right time

The first description of Functional Communication Training by Ted Carr and Mark Durand in 1985 was something of a turning point in interventions for problem behaviour because it made us think of these behaviours as being like communication¹⁵. More than this though, they showed that it was not just important to teach communication but it was also important to teach the right type of communication. In their study of five children with disabilities they first found out whether the children showed problem behaviour because it led to attention or escape from task demands. They then taught each child attention or escape gaining responses. When the child was taught the right response (for example, an attention gaining response for an attention maintained problem behaviour) problem behaviour decreased. However, when they taught the wrong response (for example, escape gaining response for an attention maintained problem behaviour) the problem behaviour stayed at the same, high level. So assessment of the reason for self-injury is important as it will tell us the most important kind of response to teach.

behaviour being shown in an extreme form. It is important to try to identify these behaviours. If they do exist they will give an important clue as to when to try to prompt someone to show a functionally communicative behaviour. Some of the behaviours that we have seen in children with Cornelia de Lange Syndrome are listed in Box 6.15 and when these behaviours are occurring this may be a very good time to prompt functionally communicative responses.

7.6.2: The effectiveness of forms of Functional Communication Training

In Section 5.3.1 we described how self-injurious behaviour can be reinforced by attention or by escape from task demands. In section 5.4 we

described how this means that self-injurious behaviour may be considered to be similar to a form of communication in that it is able to effect the behaviour of others and thus satisfy the needs of the individual. The rationale to Functional Communication Training as an intervention for self-injurious behaviour is that the communicative response that is taught will replace the self-injurious behaviour and by influencing the behaviour of others satisfy the needs of the individual. Consequently, the intervention is based on teaching a form of communication that is able to affect the behaviour of other people. This means that what is taught may be different from that which is traditionally taught as part of a speech and language therapy programme. There may be many similarities but it is important to note that the most important aspect of Functional Communication Training is the capacity of the behaviour to affect the behaviour of others.

7.6.3: Some forms of augmentative communication

In order to identify the most appropriate form of functional communication to teach, it is important to assess the child's existing method of communication. If a signing system is already in place then the task may be more one of increasing the effectiveness of responses to the communications of the child as opposed to replacing the signing system with something else. The intervention in this case consists more of ensuring that the communication is effective, i.e. is responded to by others as opposed to increasing the repertoire. Speech and Language Therapists and Clinical Psychologists can give advice on the most appropriate form of Functional Communication Training for any child or adult. Here we will describe the main types of augmentative communication that can fulfil this purpose.

Signing. There are a number of signing systems that can be taught to children and adults who have intellectual disabilities and the most popu-

lar of these is Makaton. The signing system predominantly involves the hands and consequently for some children with Cornelia de Lange Syndrome this may be problematic.

Picture Exchange Communication System (PECS). PECS is rapidly gaining popularity as a method of communication for children and adults with severe intellectual disability and/or autism¹⁷. As the name suggests the method basically rests on the child or adult either pointing to or giving pictures in exchange for a desired item or activity. There are some advantages to this method over manual signing systems. First, the child or adult can point to a picture and this may be easier for children who have upper limb abnormalities than signing. Second, one important aspect of communication is that the communication is effective i.e. that the things which somebody desires are available when they show the communicative response and thus the communicative response is reinforced. With signing it is entirely possible that the person may sign for something that is not available or that cannot immediately be made available for practical reasons. Whilst a response of "wait" or "later" may be made at this time, this may not be understood by the child and consequently the strength of that communicative response is weakened. With PECS it is possible to only make available pic-

Box 7.22: The effect of PECS on challenging behaviour

In 2002 Marjorie Charlop-Christie and her colleagues taught three boys with autism to use the Picture Exchange Communication System¹⁸. The results were good and the boys learned to use a number of pictures to make their needs known. Interestingly there was also an increase in the boys' social communicative behaviours and a decrease in problem behaviours such as tantrums, grabbing, disruption and being out of seat. Across the three boys there was a 70% or greater reduction in 10 out of 12 problem behaviours and four behaviours fell to zero levels.

tures of items or activities that can be immediately presented for any given period of time. Although this limits what is available to a child or adult for a given period of time it does mean that the pictures available will show things or activities that are possible and can be presented when requested and consequently the communicative response is further strengthened. As with Makaton there is a structured procedure for teaching PECS and recent evidence shows that it is effective for children with autism. It also shows an association with decreases in problem behaviours (see Box 7.22).

Electronic devices. There has recently been an increase in the availability of electronic devices or “Voice Boxes” that can be activated by children and adults with intellectual disabilities. These devices can be programmed to say anything that would be useful to the person. The child can then be reinforced for activating the Voice Box and thus the Box has the properties of a functional communication system (see Box 7.23). One advantage of the system over signing is that everybody can understand what is said by

the Voice Box whilst not everybody will understand all Makaton signs. PECS also has this advantage over Makaton as the word is written beneath the picture.

While signing, PECS and electronic devices are the most common forms of functional communication that can be taught to children and adults with intellectual disabilities they are not the only methods. The most important thing is to find a method that will suit the child, has a structured procedure for being taught and will be effective in all environments. We would strongly emphasise this last point. Any communication system is useless if it is not responded to. The effectiveness of Functional Communication Training in decreasing self-injurious behaviour is heavily dependent on how easily the child can reliably effect the behaviour of others in precisely the way that they want to. This key to effective intervention hinges on the idea of “Response Efficiency”.

7.6.4: The importance of response efficiency in Functional Communication Training

The technical term “response efficiency” simply refers to how effective any particular behaviour is in gaining reinforcement or reward from the environment given the cost of showing that behaviour²⁰. It is perhaps self-evident that if we have two ways of influencing the behaviour of other people then we will usually choose the way that gives us a very high return i.e. always affects the behaviour of others, and is very low cost i.e. is easy to do with no negative consequences. So, a functional communication system will generally be effective in reducing self-injury that is maintained by attention or escape from demands if the system has a higher response efficiency than self-injury. This means that when we think about teaching a functional communication system and trying to ensure that it works in the natural environment that there are number of aspects of efficiency that we must consider. This is because when the person has a choice between self-injury and functional communication they will choose the most efficient response

Box 7.23: What happens if you cannot speak?

Building on his previous work Mark Durand conducted further research on Functional Communication Training (FCT) with five children with disabilities and published his results in 1999¹⁹. This time instead of teaching the children signs or the right phrase to say he taught the to use an Introtalker that can be programmed to ‘say’ phrases using digitised speech at the press of a microswitch. Once again FCT was successful and the challenging behaviour of all five children decreased. More importantly, when the children used the Introtalker with adults who did not know anything about the programme or the children’s challenging behaviour the behaviour stayed at low levels as the adults naturally responded to the requests made by the child via the Introtalker.

Box 7.24: Responding to disruption and speech that occur for the same reason

We looked at the problem and communicative behaviours of a seven year old boy with Down's syndrome to try and understand why he might show problem behaviour when he could already communicate with speech, albeit limited²¹. We conducted some analogue experiments and watched him in his class. The results were clear. He was frequently disruptive and aggressive when asked to do tasks but would also say 'no!' and wave 'bye-'bye'. When we looked at how adults responded to these different behaviours they were much more likely to stop asking him to do something when he was aggressive and disruptive than when he said 'no!'. So his aggression and disruption were much more effective than his communication and consequently he showed these behaviours more often. The lesson here seems to be that even when someone can communicate, if it is not effective then they will show a more effective behaviour, in this case aggression and disruption.

(see Box 7.24). As this is the case we must ensure that the functional communication that the child has is much more efficient than the self-injury. Recent research has shown us that there are a number of aspects of response efficiency that are important to consider.

Low cost. Any form of functional communication must be easy to do. If the effort required to show the response is low, then the effectiveness of the system is reduced. It is important to ensure that the response that is taught is as effortless to show as is possible.

High return. It is also important that the return for any form of communication is very high and by this we mean a much higher than the return that would occur for self-injurious behaviour. This means a number of aspects of return must be attended to. First, the amount of reinforcement for any given communicative response must be

high. This means that the response that is given to, for example, a picture request for a period of attention, should be a longer period of attention than would be given in response to self-injury. Second, the response to a communicative act should be fast. This means that when a communicative response is made the reinforcer should be presented more quickly than it is presented following the self-injurious behaviour. Third, the reliability of reinforcement should be very high for the communicative response. What this means is that every communicative response should be responded to and by everybody. Whilst we recognise that it is a tall order to ask that responses to communicative acts are fast, long and reliable by rewarding the communication in this way, the communicative act becomes much more effective than self-injurious behaviour and consequently comes to replace the self-injury.

The high return for communicative acts clearly present some practical difficulties in the longer term. However, there are ways in which these may be overcome. First, there is some research evidence that it is possible to teach tolerance for reinforcer delay. What this means is that once the communicative act has been established in a child's repertoire then it is possible to signal in some way that there will be a brief period before the presentation of that reinforcer. This time period can be gradually increased from a matter of seconds to a longer period, thus allowing a more natural situation to develop. However, it is important to note that when we introduce this procedure we are necessarily decreasing one aspect of high return, that of speed. A second way in which we can overcome the practical difficulties is with the PECS system. The picture component of the system means that we can give the child or adult pictures of the reinforcing activities that are available at that time and the pictures will include only those things or activities which we know we can present quickly, for reasonably long periods and reliably. For those activities that cannot be presented in this way then the pictures can be withheld until it is practical to make them available.

For a summary of the important points about

Box 7.25: Four important things to know about Functional Communication Training (FCT) as an intervention for self-injury

1. For FCT to succeed you need to teach the right response:
 - Attention getting communication for attention rewarded self-injury;
 - ‘Stop the task’ communication for self-injury rewarded by escape from tasks.
2. Communication in FCT can take a number of forms: speech, signs, picture cards, pressing microswitches to activate tapes. The most important thing is that everyone can understand without being told.
3. Whatever the communicative response is, it should be less effort than self-injury, get more reward than self-injury, be rewarded more frequently than self-injury and get rewards faster than self-injury.
4. Be careful about communication that cannot be rewarded e.g a sign for ‘park’ when it cannot be delivered. Picture systems may be better as only pictures of things that can be done could be made available.

behaviours, such as communication, there are some general strategies that may be helpful in reducing the number of incidents of self-injurious behaviour and for which there is some research evidence. Parents and clinicians have been aware for a long while that for some individuals the lack of a structured environment can promote brief periods of anxiety in a child or adult and can lead to incidents of self-injurious and other challenging behaviours. For children and adults who find the lack of structure anxiety provoking this can be most evident at times of transition from one environment or activity to another²². This is especially the case when the child does not know which environment or activity they are going to or if that environment or activity is not the one that they expect to go to.

One way of helping with this difficulty is to try and structure the environment and daily activities using timetables such that predictability is high. This is, of course, more easily said than done. It is not always possible to run a household, a classroom or a day centre in exactly the same way each day. However, it may be possible to increase the predictability even though there is some variability. This can be done by having daily photo timetables or objects of reference available at the

Functional Communication Training see Box 7.25.

7.6.5: Facilitated communication

Whilst discussing the importance of functional communication we want to be clear about the difference between the approaches we have described above and Facilitated Communication. Facilitated Communication is a method in which a child’s hand or adult’s hand is guided by a “facilitator” to tap out messages on a keyboard. This method has been widely discredited and should not be confused with Functional Communication Training (See Box 7.26).

7.7: Additional strategies

In addition to changing the responses to self-injurious behaviour and increasing adaptive

Box 7.26: Facilitated communication

A brief and useful resource for evaluating the effectiveness of Facilitated Communication is a Fact Sheet that can be obtained from the National Autistic Society. In the Fact Sheet there is comment on a review of all the available evidence undertaken by Prof. Pat Howlin. She reviewed 45 carefully conducted studies of Facilitated Communication and found that of the 350 people who had been involved in trials of Facilitated Communication, only 6% showed any evidence of independent communication and for 90% there was evidence that the responses were unwittingly guided by the facilitators. In the United States five professional bodies have adopted a formal position opposing the acceptance of Facilitated Communication.

beginning of a day presented in the order in which the events or activities may take place. This allows the child to know what is happening throughout the day and as each event or activity becomes imminent the photograph or object of reference may be shown to the child, the child can be taken to the activity, and then the object of reference or photograph is removed. This way the child or adult may move through the day with an element of predictability. It is, of course, important to choose photographs and objects of reference carefully and to ensure that they are always associated with the activity and additionally that they are never associated with any other activity. Consistency is important to help establish the child's understanding of the relationship between a photograph or object of reference and what is about to happen.

7.8: Psychological interventions when the function of self-injurious behaviour cannot be identified

Up to this point we have indicated the kinds of interventions that the research literature indicates are effective when we are able to understand why the self-injurious behaviour is occurring on the basis of assessment and the model that we build. It is, of course, sometimes the case that we cannot work out why self-injurious behaviour is occurring and the results of the assessment may not indicate a clear cause and thus we find it difficult to select our intervention. Under these circumstances there are still interventions that we can try and again there is evidence that they can decrease self-injurious behaviour. We will deal with these interventions in the following sections.

7.8.1: A comment on punishment

The early methods of behaviour modification tended to focus on how self-injurious behaviour could be decreased as quickly as possible. Inevitably, this meant that there was a good deal of research conducted into the use of punishment as a treatment for self-injurious and other behaviour problems. The term punishment used in this

context refers to the decrease of behaviour when an aversive stimulus is presented following an instance of the behaviour. Following heated debates in the late 1980s and early 1990s the description of punishment techniques in the research literature has decreased and has been replaced by the approach which we have adopted in this book. That is, an approach which tries to uncover the reasons for self-injurious behaviour, replace the behaviour with a more adaptive response and manage the behaviour in a way that would lead to its eventual decrease. Whilst this is the approach that is now advocated by many researchers and clinicians in the field, there is still the possibility that punishment can be proposed as a method of control of the self-injurious behaviour of people with intellectual disabilities. In this section we want to draw attention to the three main issues that are important to consider.

First, punishment tends to give a short term success. There is some evidence in the research literature that the behaviours decrease more quickly when punishment methods are used than when alternative approaches are adopted, although there is some debate about whether this is the case. One factor that may be related to the short term success is that the punitive stimulus delivered, needs to be quite severe or unpleasant in order to suppress the behaviour. Inevitably this may lead to the use of punishers in a way that may be considered inhumane. For example, squirting lemon juice into the mouth, enforcing physical activity and inducing pain. Whilst there is evidence that these procedures may lead to short-term success there is also evidence that when children and adults who have been involved in these treatments are followed up at a later date the self-injury has returned and persists²³.

Second, whilst punishment may decrease self-injurious behaviour, it does so simply by teaching somebody what not to do. However, if we accept the model that is described in Chapter 5 that the behaviour is functional then if we use punishment alone, clearly we do not teach the person what to do instead of self-injuring. This means

that the person will have the same needs but no way of satisfying those needs. If the model is correct then this means the self-injury will occur again at a later stage and the evidence on punishment shows that this is exactly what happens.

The model that we have presented in Chapter 5 shows how self-injurious behaviour can be learned. Punishment as an effective treatment also depends on learning i.e. the person learns not to self-injure because the punishment will follow. If this is the case then there is agreement that the self-injurious behaviour is learned. The question then is how best to affect the unlearning of self-injurious behaviour. Our argument is that if there is agreement that self-injurious behaviour is a learned behaviour then either approach may work but we would argue that using positive methods is preferable to punishment simply because of the short term success issue, the inhumane nature of many punishment methods and the importance of replacing the behaviour with a different behaviour.

This discussion of punishment presents a brief summary of our views and we accept that there are different opinions on the use of punishment as an intervention for self-injurious behaviour. Clearly, it is a personal decision as to whether punishment techniques are used and not all punishment techniques will necessarily be inhumane. If you are considering using punishment techniques to decrease self-injurious behaviour we would strongly advise you to seek advice from a clinical psychologist or behaviour analyst and to keep all of the effects of punishment under close review.

7.8.2: Differential reinforcement

One strategy for decreasing self-injurious behaviour for which there is good evidence is differential reinforcement. This simply means presenting some kind of reward when an undesirable behaviour is not occurring and thus trying to make the consequences of not self-injuring “better” than the consequences of self-injuring. There are a number of ways in which this can be carried

out and these are described below. However, before describing the various methods of differential reinforcement it is important to know some of the basic principles that underlie how this is thought to work.

For sometime it was believed that differential reinforcement worked because it was rewarding the person for not doing something as opposed to doing something. However, it now seems likely that it may work in a different way and is effective because the reward that usually follows self-injurious behaviour is now presented at a different time and consequently the person showing self-injurious behaviour does not need to show self-injury in order to gain the reward. If you look back at the model described in section 5.3 and think about a child showing self-injurious behaviour in order to gain attention then that if we present attention at regular intervals then it makes it more likely that the child will not need to self-injure in order to gain the attention, as the attention is occurring with sufficient regularity.

One of the problems with some forms of differential reinforcement is that, a bit like punishment, it does not necessarily teach the person what to do. So, if we present attention as a reward for not self-injuring then the person may not have shown any particular behaviour that would normally gain attention. All they are doing is not self-injuring. This means that the person has not learned a behaviour that they can use in the future to gain attention. For this reason Functional Communication Training, discussed in section 7.6, tends to be favoured over differential reinforcement techniques. However, when we are unable to identify the reinforcement for self-injurious behaviour, and thus the function of the behaviour, differential reinforcement can be a useful strategy to try to decrease self-injury.

7.8.2.1: Important aspects of differential reinforcement

There are four aspects of differential reinforcement that are important to consider before starting any kind of programme.

Reinforcer selection. If you are going to present a reward for no self-injury occurring for a given period of time then clearly it is important that the reward you choose is indeed a reward for the person. Different people have very different preferences in terms of what they find rewarding. Before starting the programme it is important to draw up a list of things that the person finds rewarding and to ensure that more than one type of reward is available.

Avoiding satiation. One of the problems with presenting a reward after a period of no self-injury when the period is very brief, is that people may very quickly satiate to the reward. What this means is that when you have had too much of a good thing then you do not want any more! Three ways of dealing with this problem are to use very brief amounts of rewards, to use a variety of different rewards and to use symbolic rewards, such as stars on a chart that can be exchanged for an item of the person's choice.

Consistency. When using any reward programme it is extremely important that there is a high degree of consistency. This means that when the reward system is set up the reward is always delivered after a period of no self-injury, for example, and this is carried out by everybody in all environments.

Changing the programme over time. In the initial stages of any differential reinforcement programme the time period for which a behaviour should not occur needs to be fairly short so that the person has a very good chance of success. One way in which the time period can be calculated is by working out the average period of time that elapses between incidents of self-injurious behaviour and then dividing this by three. So, if self-injurious behaviour is seen on average every 15 minutes, then the initial differential reinforcement period would be 5 minutes and the reinforcement would be delivered if there is no self-injury for a 5 minute period. Also, in the early stage of the programme the amount of reinforcement

that is delivered should be high for a short period e.g. 2 minutes of attention for 10 minutes of no self-injury. Over time, as the programme begins to be effective it is important to increase gradually the amount of time that the person does not show self-injury for (10 minutes, then 15, then 20 etc.) and to gradually decrease the amount of reinforcement that is given (2 minutes of attention then 1, then 30 seconds etc.) in order to avoid satiation which we discussed above.

7.8.2.2: Types of differential reinforcement

There are three basic types of differential reinforcement.

Differential reinforcement of other behaviour (DRO). DRO means presenting reward when a behaviour that has not occurred for a predetermined period of time. Thus, it does not matter what happens during the period of time, the person can be doing anything except self-injury, what matters is that they did not self-injure. If the person does self-injure then the time period starts again.

Differential reinforcement of incompatible behaviour (DRI). DRI is similar to DRO but in this case the reward is presented when the per-

Box 7.27: The importance of rewarding incompatible behaviours

An old study but an important point. In 1974 James Young and John Wincze described their intervention for head banging and hitting for a 21 year old woman with profound intellectual disability²⁴. They showed that if they presented a reward if the woman kept her hands on an object then head hitting decreased but head banging increased. This shows us two important things. The first is that rewarding an incompatible behaviour can work, hands on an object is incompatible with head hitting. The second is that if someone has not been taught a behaviour to replace the self-injury, such as communication, then another behaviour may increase.

son has shown a behaviour that is incompatible with self-injurious behaviour for a given period of time. So, if it is decided that a DRI programme will be implemented for head hitting then a child may be reinforced for playing with toys with his hands for a predetermined period of time because the behaviour of playing with toys with hands will be incompatible with head hitting. If the person does self-injure then the time period starts again. There is some limited evidence that DRI is more effective than DRO (see Box 7.27).

Differential reinforcement of alternative behaviour (DRA). We have already discussed one form of DRA and that is Functional Communication Training. Differential reinforcement of alternative behaviour means presenting reinforcement for a behaviour that has the same function as self-injurious behaviour. So, if a child shows self-injurious behaviour in response to a task demand then the DRA programme would consist of terminating the task demand when the child shows a behaviour such as making the sign for “break” as opposed to showing self-injurious behaviour. The sign for break would be the alternative communicative behaviour.

7.8.2.3: Ways in which differential reinforcement can be delivered

There are three basic ways in which the differential reinforcement can be delivered.

Whole Interval Method. This method would be used when running a DRO or DRI programme. The method entails watching the child for the all of the period that you have set in the programme. So if the programme consists of DRO after five minutes, then you would watch the child for five minutes and if the behaviour has not occurred then the reinforcer is presented. If the self-injury does occur then the time starts again. The obvious problem with this method is that it means constant observation of the child in order to ensure that the behaviour has not occurred for five minutes. Inevitably, this has led

to problems with implementing this approach but thankfully there are two other ways in which the differential reinforcement programme can be implemented.

Momentary DRO. This method is much easier to implement than the whole interval method and consists of setting a period and then only observing the child at the end of that period and presenting the reward if the child is not self-injuring when that observation takes place. So, if the programme consists of a Momentary DRO (or DRI) with a ten-minute interval, then running the programme comprises having a watch with a count-down function that will bleep after 10 minutes. When the watch bleeps, the person running the programme will briefly look at the child and if the child is not self-injuring then the reward is presented. If the child is self-injuring then the reward will not be presented and when the self-injury has stopped the time will be reset and the time period starts again (see Box 7.28).

Noncontingent reinforcement (NCR). This is perhaps the easiest way of presenting reinforcement but it is debatable as to whether this is a method of differential reinforcement or whether it works in a different way. To take the example of a child showing self-injurious behaviour who finds atten-

Box 7.28: Comparing methods of differential reinforcement

Alan Repp and his colleagues compared two different methods of differential reinforcement to try to decrease the problem behaviour of three 7 year old boys with intellectual disabilities²⁵. They found that the whole interval method (in which a reward was gained for not showing problem behaviour for 5 minutes) was more effective than the momentary method (in which a reward was gained if for a brief period at the end of a 5 minute interval) the child was not showing problem behaviour. Importantly they also found that if the momentary method was used after the whole interval method than it was just as effective.

Box 7.29: Noncontingent reinforcement versus DRO

For many years there was a strongly held view that one of the best ‘positive’ interventions for self-injury was DRO (presenting a reward or reinforcement if self-injury has not occurred for a period of time). In 1993 Timothy Vollmer and his colleagues published a report showing that noncontingent reinforcement was equally effective²⁶. This means that if you know the reinforcer for a behaviour (say attention) then instead of running a DRO programme (where you have to watch for, say, 10 minutes and if the behaviour has not occurred then present the reward of attention) you could try noncontingent reinforcement and present the reward of attention every tenth minute regardless of what is happening. This finding really needs to be replicated but for the time being it shows us that there is a more practical alternative to DRO.

tion reinforcing, noncontingent reinforcement means setting a time period and then presenting a period of attention after that time period has elapsed regardless of whether or not the self-injurious behaviour has occurred. This sounds very curious as we have previously argued that it is important not to present reinforcement when self-injurious behaviour occurs. However, there is some research evidence that using this method is as effective as differential reinforcement methods for some children (see Box 7.29).

Noncontingent reinforcement may work by simply ensuring that the child receives attention at regular intervals thus meaning that the child does not have to self-injure in order to gain the reward. If this is the case then it is important that the interval is very brief and that the programme is rigorously maintained. The advantage of this method over the whole interval method is that for the whole interval method it is preferable if there is no response to the self-injurious behaviour. However, this may lead to extinction occurring with the inherent problem of increased injury. Using this method may offset the problem of extinction whilst being able to run a much

more manageable programme than that described in whole interval methods.

The above descriptions may appear somewhat bewildering given the variations that could be used. Our advice is as follows. The best alternative would be DRA in the form of Functional Communication Training. However, we recognise that it may not always be possible to identify the function of behaviour consequently it is not easy to put this kind of programme into practice. We cannot teach a behaviour as an alternative way of satisfying a need if we do not know what the need is. So, the next best method would be the whole interval method followed by Momentary DRI with the incompatible behaviour being an adaptive behaviour that is likely to be intrinsically reinforcing for the child. However, in order to offset the problem of extinction it is necessary to ensure that there is a response to the self-injurious behaviour when it occurs that ensures the safety of the child. This will commonly mean some kind of physical restraint by others until the self-injury abates. During this period it is absolutely critical that the response of the person involved in the restraint and the restraint itself is not experienced as reinforcing or rewarding by the child. This means that the response of the adult must be cool and should not include eye-contact or talking to the child as these may be rewarding. How rewarding this is will differ depending on the child and it is important that the response of the child to this approach is monitored and records are kept to evaluate the effectiveness of the approach. Finally, we would strongly advise seeking advice from a clinical psychologist or behaviour analyst before implementing any restraint procedure

The second piece of advice that we would give is that differential reinforcement procedures and NCR should be seen as helping to gain some decrease in the self-injurious behaviour and giving a window of opportunity for increasing the child’s communicative behaviours in the ways that we have described in section 7.6. So, it may not be a procedure that would have to be used for a very long period of time. There is no doubt

that the procedure is hard work and requires a good deal of input. This may be made easier by gradually lengthening the period of time between rewards whilst keeping a close eye on the effectiveness of the programme.

7.9: Exercise

Within the last decade there has been a small number of research papers that have described a beneficial effect of physical exercise on self-injurious and stereotyped behaviours. Whilst it is not clear why exercise should decrease self-injurious behaviour the effect on some people is notable. We do not underestimate the difficulty in trying to find physical exercise that people may find enjoyable and would be willing to participate in and, of course, some people will be limited by physical disability. Nevertheless, for the sake of completeness we should note that there is evidence that this intervention may be effective.

7.10: Generalisation and maintenance

There are two issues associated with behavioural interventions for self-injurious behaviour that have caused both clinicians and researchers significant challenges over the past 30 years. First, whilst it is possible to decrease self-injurious behaviour in one environment using a fairly structured programme the gains do not necessarily naturally occur in all environments. In other words the decrease in self-injurious behaviour can often be specific to the place in which the programme was first conducted and the people who were there. The second issue is maintenance. This means trying to ensure that any early gains in decreasing self-injurious behaviour are maintained over time. Whilst reading this chapter you have no doubt been struck by the amount of planning and effort that goes in to running a successful behavioural programme. It is perhaps inevitable that over time it becomes more and more difficult to keep these intensive programmes running. We will deal with the issues of generalisation and maintenance in turn.

7.10.1: Generalisation

One way in which generalisation can be assisted is by carefully programming the gradual implementation of a programme into all environments. So, if a behavioural programme is shown to be successful in one environment then it can be taken into new environments in a systematic way. In practice, this means sharing information with others and may also mean convincing others that the intervention is worth pursuing. This can be helped if there are records kept of the effectiveness of an intervention so that others can see that an approach is successful. Another way in which this kind of generalisation can be promoted is by the person who has been closely involved with the programme in the early stages going into the second environment and working alongside those who will be taking the programme over. This pairing of the person who initially runs the programme and those who will be running the programme in the new environment could also take place where the programme has already been running.

The second way of trying to promote generalisation, and also may be helpful in reducing the onerous nature of a programme, is to use a novel and clear signal that identifies the programme as running and thus indicates to the child or adult that a new learning process is in operation. In practice what this means is that when a programme is first implemented the clear and novel signal is placed in an obvious position and the child is clearly aware that the signal is present. The signal can be anything that is new, and thus will only be associated with the programme, portable, and thus can move with the child wherever the programme and the child need to go, and highly visible to the child. The signal can take any form such as a brightly coloured large card with a unique design on it or an item of clothing that has never been worn before and so is always visible to the child.

The signal should then be shown whenever the programme is running but should never be shown when the programme is not running. This

way the person learns to discriminate between those times when the new learning is taking place and those times when it is not. This may be helpful in two ways. First, there are always times when it is simply not possible to run the programme and this gives us a bit of breathing space without losing the effectiveness of the programme. The second is that when we come to generalise the programme to a new person or a new environment we can take the signal with the child and this will help the child to know that the new learning is now operating with the new person or environment.

7.10.2: Maintenance

Maintenance of programmes over time is particularly problematic. One of the things that we have become aware of is that when the self-injurious behaviour has decreased somewhat and things are not so problematic there is a tendency for the behavioural programmes to be used with less consistency because there is less urgency. The problem then is that the behaviour may return as the new learning becomes undone. This means that ensuring the consistency of programmes over time is extremely important. There are two issues that might help with maintenance. The first is that whatever the programme implemented, it needs to be as minimally demanding of others as is possible whilst still being effective. This means, for example, trying to increase the period of differential reinforcement programmes as rapidly as possible while still maintaining any gains. Secondly, we believe it is extremely important, and there is research evidence to support this, that an adaptive behaviour that can replace the self-injurious behaviour is taught to the child or adult at an early stage in any programme. There is good evidence that Functional Communication Training can maintain overtime because of its inherent capacity to give the child control over their social environment. In other words there is natural maintenance in programmes that manage to build up children's adaptive behavioural repertoires.

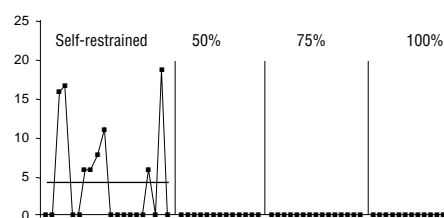
7.11 Loss of control

For some children and adults increases in self-injurious behaviour in both frequency and intensity may be unrelated to environmental events or medical causes such that it appears that they have started to lose control over the behaviour. This idea is somewhat contentious but we feel that there are some aspects of self-injurious behaviour that should lead us to think that the person may not have complete control over the behaviour. The first is the presence of self-restraint or preference for imposed restraint. We have discussed these in Chapter 5. The second is the association between self-injurious behaviour and compulsive behaviours and, to a lesser extent, hyperactivity and stereotyped behaviour. These associations make us think that the person has difficulty in inhibiting their own behaviour. A lack of behavioural inhibition, this means not being able to prevent a behaviour from starting or not being able to stop a behaviour once it has started, is a feature of hyperactivity and compulsive disorder. As we discussed in Chapter 6 it may well be that self-restraint and preferred imposed restraint are indicators that the individual is unable to inhibit their behaviour and consequently needs to do so physically.

7.11.1: Using protective devices

When behaviours occur at a higher level of frequency and intensity there is a temptation to use protective devices such as arm splints, gloves, and helmets in order to limit the amount of self-injury and the consequent damage. There are a number of issues associated with the use of protective devices that it is important to consider. First, using the devices such as padded gloves might help with ignoring the behaviour as part of an extinction programme (see above) because the chance of injury is reduced. However, we have noted the importance of not conducting an extinction programme in isolation because the person might easily develop another form of the behaviour and then there has been no gain. So,

Box 7.30 Replacing self-restraint with protective devices that can be faded



We carried out an intervention for Dawn, a 25 year old woman who had profound intellectual disability and vision and hearing impairments²⁷. She punched her head and body and self-restrained by wrapping her hands tightly in clothes and pushed her hands into wooden objects. As can be seen in the graph although she self-restrained she still punched her head and body about 5% of the time. We then introduced arm splints which had an adjustable joint on the elbow. At first we set the degree of movement to about 50% of the full range. During this period Dawn did not self-injure at all as the head and body punching were restricted by the splint. After a time we increased the range of movement to 75% and then 100% (full, normal range of movement) without self-injury occurring. We were able to put the splint under Dawn's clothing so that it did not appear odd and whilst wearing the splint she was less restricted than when she self-restrained.

before using protective devices it is extremely important to assess the function of self-injurious behaviour as we have described in Chapter 6.

The second issue is that the use of any protective device can give rise to physical problems such as irritation of the skin and, with straight-arm splints, atrophy of the muscles. It is important therefore to ensure that medical advice is sought both prior to and during the use of protective devices. The third issue is that if the person is unable to inhibit their behaviour then whilst protective devices may help them to do so in the short term they may come to rely heavily on the presence of the protective devices in

order to control the self-injurious behaviour. Thus, a preference for this imposed restraint may develop and it is important to plan how the restraint may be faded over time in order to avoid a high level of constant restraint.

This kind of intervention i.e. the gradual fading of preferred imposed restraints, can be very successful. There is, curiously, an advantage in an individual having a high preference for imposed restraint. First, it usually means that the behaviour is under control. Second, it can mean that the restrictive nature of the device can be reduced over time without losing the control over the self-injurious behaviour. We have managed to achieve this in the past in people who have a preference for wearing arm splints to control their head-punching. We were able to gradually introduce more movement into the elbow joint of straight-arm splints and also gradually reduce the length of the straight-arm splints, down to a cuff around the wrists, without losing the control of the protective device. (See Box 7.30). This kind of intervention is difficult to implement and we would strongly advise seeking the advice of a clinical psychologist or behaviour analyst before trying to implement the intervention.

The use of protective devices is clearly contentious as it may be seen as a punitive method of intervening with self-injurious behaviour. However, protective devices are often used simply out of the desperation of a parent or carer to protect the person they care for from injury. We would strongly advise that before using protective devices carers should seek the advice of a clinical psychologist or behaviour analyst and the advice of an occupational therapist or physiotherapists to ensure that the device can be faded over time. Our experience is that it may take some time to find the right device for an individual but if the device does gain control over very severe and intense self-injurious behaviour then the fading process can be effective. Finally, we would urge extreme caution in using devices when there is a clear social function to the behaviour. Under these circumstances the effect may be very similar in some ways to that of pun-

ishment and extinction in that all we have done is suppress the behaviour of the individual and not given them a different way of meeting their needs. Under these circumstances we would expect the behaviour to return or another form of the behaviour to occur.

We noted in the introduction to this chapter that it is important to ensure that wounds heal as quickly as possible. We do recognise that this is easier said than done, nevertheless it is important to try to pursue any strategy to promote healing. One way in which this can be done is by ensuring that the site of injury is covered. It may take some imagination in order to find the right way of keeping a wound covered but it is worth the effort. This seems to help by promoting healing so getting through the scratch-itch cycle that accompanies healing more quickly and thus avoiding constant scratching that leads to further injury and so on. Additionally, we have noticed that some people who show self-injurious behaviour do deliberately cover up the site of the injury and we believe that this may be a self management strategy that helps people to inhibit the self-injurious response. That is by removing the visual stimulus of the wound, the self-injury seems to occur less. Some of the issues that associate to covering a site of self-injury are those which we have mentioned with regard to protective devices. It does appear that some people come to rely on a wound being covered and will prefer to have bandages, for example, covering their wounds. If this is the case then it may be possible to fade the size of the bandage over time to become a symbolic form of control.

7.11.2: Managing and changing self-restraint

Our past research has shown that self-restraint does appear to be common in children and adults with Cornelia de Lange Syndrome who show self-injurious behaviour. As we have mentioned previously we think this indicates that there may be some lack of behavioural inhibition and thus the behaviour is difficult for the person to control. As we noted in Chapter 6 self-restraint

can take many forms and can be restrictive for the individual to a greater or lesser degree. If self-restraint does not appear to prevent the individual from taking part in activities and there are no physical consequences to self-restraint, then it may be it the best strategy to allow the self-restraint to continue but to ensure that it does not become more restrictive. However, when self-restraint impairs the individual and may be causing physical harm, for example some people can wind their hands in their clothes so tightly that circulation of the blood is compromised, then it is important to try and reduce the amount of restraint while still maintaining control over the self-injurious behaviour.

The most important thing in changing self-restraint is not to lose the control over the self-injurious behaviour. In some ways the self-restraint is a real asset in that it goes everywhere with the person, it is effective and it is under the

Box 7.31: Finding the right form of protective device before fading

In Box 7.30 we described how we were able to introduce a new splint for Dawn to control head punching and then gradually increase the amount of flexion at the elbow without the self-injury increasing. However, we did not get it right first time. The first splint we introduced finished at the cuff. When we tried this with Dawn she became distressed and anxious, tried to hit her head and broke the elbow joint within two minutes. The next splint we tried had a band extending from the cuff, over the back of the hand between the thumb and first finger and back to the cuff. Dawn preferred this and was calm when the splint was put on and would hold out her arm to help. In hindsight we should have looked more closely at how Dawn self-restrained. If we had done so we would have seen that she always had either clothes or something else over the back of her hand and running between her thumb and first finger. This was the most important part of the self-restraint for Dawn and we failed to build it into our first splint.

person's control. The trick is to try and reduce the amount of restriction the person is experiencing without losing control. There are a number of research papers and descriptions in the literature of how self-restraint can be decreased to a symbolic level whilst still keeping control over the self-injury²⁸. In order to do this effectively it is important to try to identify what aspect of self-restraint the person usually prefers. So, for someone who likes to wind their hands in their clothes, it could be that they prefer the tight sensation around the wrists, it could be the precise site of the restraint e.g. around the arm, or the type of restriction that is experienced e.g. total movement restriction or just restriction of the lower arm. Once it has been possible to identify the aspect of self-restraint that a person really needs then other facets of the restraint can be faded i.e. gradually reduced over time whilst leaving the important stimulus intact. (see Box 7.31).

For a summary of the main points about the use of protective devices see Box 7.32.

7.12: Medication

It is beyond the scope of this book for us to discuss medications that might be used in order to decrease self-injurious behaviour but we can give some indication of the current thinking on this topic. You will remember that in Chapter 3 we discussed disorders of neurotransmitters that might be related to self-injurious behaviour. The three types of neurotransmitters were opiateergic, serotonergic and dopaminergic. Generally speaking medication that has been associated with decreases in self-injurious behaviour tends to target disorders in these neurotransmitters. However, we should note that in the research literature that there is very little evidence that medications that have these actions are effective for people with intellectual disability. There have been very few large scale trials and the evidence that exists tends to be descriptions of single cases. Many authors writing on the topic of medication for self-injurious behaviour conclude that the evidence is more suggestive than conclusive

Box 7.32: Four important things about using protective devices

1. **Do not use protective devices as the only form of intervention. They should be combined with a behavioural programme.**
2. **Before using devices seek medical and psychological advice. Be aware of irritation to the skin and other problems such as muscle atrophy (wasting).**
3. **It is possible that people will become 'addicted' to their protective device. Before introducing the device develop a plan about how the device can be faded.**
4. **If people begin to like their devices (hold their arms out, 'ask' for the device) they can reward self-injury if they are used after self-injury has occurred.**

and we would agree with this position. At present therefore, it cannot be said that there is a medication available that can be used for all people with self-injurious behaviour. Further information on possible medications that can be used for self-injurious behaviour is given in Dr. Tom Gaultieri's book that is listed in the Bibliography and discussion of medications can be found on the Cornelia de Lange Syndrome website.

7.13: Summary

Interventions for self-injury should be selected on the basis of the cause of self-injury. Medical interventions for conditions that give rise to pain and discomfort should be implemented first. For self-injury that occurs because of the resultant stimulation, intervention comprises both reducing the stimulation and presenting competing stimulation. For self-injury that occurs because of social rewards, the intervention should comprise modifying the response to self-injury and teaching alternative responses to displace the self-injury, Functional Communication Training is the favoured approach. For self-injury that does not appear to be maintained by sensory or social

rewards, differential reinforcement may be effective. Protective devices should be used as a last resort with thought given to how they will eventually be reduced. Any intervention should be evaluated with records kept of the levels of self-injury before the intervention and whilst it is being conducted. For any intervention, advice from a Clinical Psychologist or behaviour analyst should be sought.

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Chapter 8: Conclusions

8.1: Prevention

It is often said that prevention is better than cure and there is little doubt that this applies to self-injurious behaviour and any other behaviour problems. The most important thing that carers can do in order to try to prevent self-injury from developing is to be aware of the possible causes of self-injurious behaviour and the theories of why self-injurious behaviour can develop. It is important therefore, to be aware of the information and models that we have presented in Chapter 2 and Chapter 5.

More specifically there are some things that we would recommend that parents of children with Cornelia de Lange Syndrome attend to. The first is that a careful eye is kept on children with Cornelia de Lange Syndrome with regard to behaviours that might later turn into more severe self-injurious behaviour. This means trying to be alert to what can be a very mild behaviours such as gentle hand biting, soft head-banging and gentle scratching. These behaviours are sometimes referred to as proto-self-injurious behaviour in that they are thought to later develop into more severe self-injurious behaviour. It is also important that others who are involved with the child are made aware of the potential for self-injury and they should be alert to the early signs of these behaviours. Communication is clearly important at this stage and using diaries that travel with the child between different environments will help to ensure that everyone is looking for the same thing and aware of how to respond should it occur.

If these early behaviours are identified then the most important thing to do is to work through the possible causes we have identified

in Chapter 5 and check your response, and the responses of others who are in contact with the child, to these behaviours. By this we mean ensuring that your responses to these behaviours are not rewarding and thus the social reinforcement that we have described in Chapter 5 does not become operative. It is important to remember that your natural response to any self-injurious behaviour that is shown by your child will be to comfort and protect your child. However, it is also important to remember that this kind of natural response can be experienced by the child as a reward and consequently the behaviour will increase in the future. Understanding how and why this happens will be important in trying to prevent the behaviour from increasing over time. The future development of self-injurious behaviour in children and adults with Cornelia de Lange Syndrome, and in fact in any children with intellectual disability, is not inevitable. By checking early responses to the behaviour it is possible to decrease the chances that the behaviour will increase over time.

As we noted in Chapters 2 and 5 we believe that pain and discomfort are related to self-injurious behaviour in Cornelia de Lange Syndrome and anyone who has an intellectual disability. It is important therefore that any pain or discomfort that is experienced by the individual is dealt with as quickly as possible in order to reduce the chance that self-injurious behaviour may occur and may then become socially rewarded and thus increase over time. Being aware of the types of health problems that children and adults with Cornelia de Lange Syndrome experience, and the types of intervention that need to

be brought to the attention of health professionals are important aspects of preventing self-injurious behaviour from developing. It is important that when pain and discomfort are present and that you believe mild self-injurious behaviour is occurring in response to pain and discomfort (Chapters 2, 5 and 6) that you act quickly, inform health professionals and seek effective intervention. Further information on health problems associated with Cornelia de Lange Syndrome can be found on the Cornelia de Lange Syndrome website.

Should self-injurious behaviour begin to develop then there is no harm in seeking early advice from a clinical psychologist or behaviour analysts. In the early stages intensive intervention may not be necessary and some advice from professionals who are familiar with self-injurious behaviour may go a long way. It made help to have advice from someone on how to respond to self-injurious behaviour when it occurs in the early stages and to be able to plan for the future.

The final aspect of prevention that we believe is extremely important in preventing self-injurious behaviour from developing into a socially reinforced and thus functional way of interacting with others, is the development of an effective and reliable communication system for the child. We cannot emphasise enough how important we believe it is that every child, regardless of the risk of developing self-injury, is able to make their needs known to others. There is very good evidence that children with all degrees of intellectual disability can develop basic but effective communication systems, and there are a variety of ways in which this can be achieved. We are also aware that it is difficult to find external help to teach effective communication systems and our experience is that those parents who lobby hard for this are those who are more successful in finding help. For children with a greater degree of intellectual disability, signing systems such as Makaton are effective and there is increasing evidence that PECS (Picture Exchange Communication System) is useful in reducing behaviour problems generally.

8.2: Assessing and intervening

Finally, there are some general points that we would make about assessing and intervening when self-injurious behaviour is occurring. First, it is hard to take on self-injurious behaviour on your own. One role for carers is being aware of the various parts of an intervention that are necessary and trying to bring together people who have contact with the child to work towards the same goal. Additionally, it is important to include in the team outside help from clinical psychologists, behaviour analysts, speech and language therapists and others who can make an important contribution. As we have said at various points throughout the book, our experience is that parents who lobby are more successful in bringing together these groups of people than those who do not.

We have emphasised the importance of building a model of the causes of self-injurious behaviour for each person and then implementing an intervention that is based on this model. We believe that this is the most effective way of approaching the problem and putting together an intervention that is likely to be successful in different environments and over time. However, we have acknowledged that it is not always possible for assessments to show us the causes of self-injurious behaviour and then we fall back on a systematic trial-and-error process in which we may try various forms of behavioural intervention in order to find that which keeps the behaviour at a low level. There is nothing wrong with a trial-and-error approach. Whatever intervention is employed the most important thing is that accurate records of the self-injurious behaviour are kept in order to evaluate whether or not the intervention is reducing the behaviour. These records will help decision making in the short term but will also help in the longer term in being able to look back at what was effective and when it is necessary to return to an intervention.

Finally, there are three things which we believe are important in trying to get interventions to work. The first is that there is agreement amongst all the carers who are involved with a

child or adult showing self-injurious behaviour about the intervention that will be conducted and how and when it will take place. This is, of course, easier said than done and it may well mean that some degree of compromise will be necessary in order to achieve agreement. However, if there is no agreement then the evaluation of an intervention will not be possible as we cannot be sure precisely what is being done. Second, consistency in an intervention is critical in order to ensure that the intervention has been given a good trial. This means people who are involved with a child or adult agreeing to run an intervention in the same way for whatever time period has been agreed. Third, help with running an intervention is extremely important. Any behavioural programme is unlikely to show an immediate effect and commonly the interventions take time to show a reduction in self-injurious behaviour. It is important to persist both with individual interventions but also to be prepared to try another intervention if one simply does not work.

8.3: When interventions fail

It is worth thinking about why interventions fail if they do. This is important both because there may be things about the intervention that we can change but also because failures can often tell us something about why the self-injurious behaviour is occurring. Any failed intervention (which means that when we review the records there appears to be no decrease in self-injurious behaviour or there has been an increase) should be reviewed to try to uncover the reasons for failure.

There may be a number of reasons why interventions are failing or appear to fail. The first is that the assessment has not identified the right cause of the self-injurious behaviour and consequently the wrong type of intervention was put into place. (See Box 7.21). When this happens it is important not to dismiss behavioural interventions as not working, rather it is important to understand that getting the assessment right and identifying the cause will help with putting together the right intervention. The second is that

the extinction burst referred to in box 7.13 may be taking place. You will remember that this means that when we first stop presenting a reward for a behaviour, the behaviour can temporarily increase in frequency and intensity prior to decreasing. This means that if we see this early increase in intensity and frequency, it may not be that the intervention is failing rather that it is the right intervention but that we are seeing an extinction burst.

A third reason is that the intervention may simply not be implemented across the board. There are two ways in which this might be apparent. The first is that the records may show that the intervention is working in one environment but not elsewhere. Under these circumstances it is important to look at the two environments and try to work out whether the intervention is being implemented in exactly the same way in both environments. The second is that an intervention that has worked in the past does not seem to work now. We noted above that it is difficult to sustain interventions over time and that when self-injury starts to decrease there is a temptation to take a bit of a breather. It is important to keep programmes under review and ensure that they are being carried out with persistence.

In addition to these specific reasons we do believe that the causes of self-injurious behaviour can differ over time and that what is causing self-injurious behaviour at one point in time may not necessarily be important later on. If interventions stop working therefore, it is important to go back to re-assess the potential causes of self-injurious behaviour as another cause may have emerged over time. Similarly, just because an intervention does not work at a given time does not necessarily mean that it will never work. If the intervention was not correctly matched to a given cause then that would be the reason why it did not work. If the cause does become influential later on then the intervention that previously failed may now be effective. The message is do not throw out interventions completely, they may work at a different point in time. Again, record keeping will help us to decide.

8.4: The goal

The final thought we leave you with is that it is possible to decrease self-injurious behaviour. The key to doing so is to adopt a systematic approach to assessment and intervention and to be guided by the records that you keep of the results of intervention. The first intervention you try may not work, the important thing is to keep trying

and to persist with different kinds of intervention based on what you believe the causes are, keeping records to tell you when you are beginning to succeed. We firmly believe that with persistence, resources and time it is possible to reduce self-injury in all children and adults with Cornelia de Lange Syndrome to a level that ensures safety and does not significantly impinge on quality of life. That is the goal.

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