



**ADULTS WITH WILLIAMS SYNDROME GUIDELINES  
FOR FAMILIES AND PROFESSIONALS**

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## **FOREWORD**

Williams Syndrome (WS) is a rare condition, occurring in approximately 1 in 20,000 live births. As a result, most people will not have encountered an individual with WS before, and they will understandably know little, if anything, about the characteristics and difficulties associated with this condition. The purpose of this booklet is to provide information about this rare disorder, and to outline the difficulties that may be experienced by adults with WS. Of course every individual is different, and not everyone with WS shows all of the characteristics and difficulties described below. The contents of this booklet are based on research undertaken over the last 25 years. This research demonstrates that WS is associated with a number of distinctive behavioural and psychological features, which differentiate individuals with this condition from other groups of adults with learning difficulties. These characteristics, and their implications for the supported management of adults with WS, are discussed in detail throughout this booklet. It is hoped that the advice and suggestions that follow will be of benefit to those who live and work with adults with WS. A separate information sheet for employers and work supervisors can be obtained from the WS Foundation, and families may choose to forward that document to potential employers and supervisors who may not have time to read this more detailed booklet.

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Published by:

The Williams Syndrome Foundation

161 High Street

Tonbridge, Kent TN9 1BX

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## GENETIC BASIS OF WILLIAMS SYNDROME

Williams Syndrome is a genetically determined intellectually disabling condition. Williams syndrome is a rare genetic disorder, with a prevalence of between 1 in 7,500 and 1 in 20,000. It is caused by a deletion of approximately 21 genes on one copy of chromosome 7. This is sometimes known as the Williams syndrome critical region (WSCR). Our understanding of the consequences of the deletion of these genes is very limited. However, research is beginning to identify the effects of some of the commonly deleted genes. The first of the deleted genes identified was the elastin gene. Elastin is an important constituent of connective tissue in the body (including blood vessels, muscles, ligaments, skin), and reduced or abnormal elastin may explain some of the physical and medical characteristics, as well as the distinctive facial features (see photographs), associated with WS. The deletion of the elastin gene is extremely common in WS, occurring in 95-99% of individuals. As such, it is often used as a genetic marker for WS, with the deletion typically being assessed using Fluorescent in-situ Hybridisation (FISH). Other deleted genes have been hypothesized to have roles in visuo-spatial difficulties (LIMK1), level of intellectual disability (GTF21), facial characteristics (GTF2IRD1), differences in brain structure (CYLN2), hypercalcaemia (BAZ1B) and diabetes (STX1A). Overall, it appears that while some genes may have a direct effect, many genes may interact with other genes at various stages throughout life to produce the characteristics of WS. Although most cases occur sporadically, identical twins have been reported, and there are a few accounts of parent to child transmission.

## PHYSICAL AND MEDICAL FEATURES OF WILLIAMS SYNDROME IN ADULTHOOD

Individuals with WS are often of short stature: the average adult height is 5ft for females and 5ft 6in for males. Although many children with WS are of slight build, there is a tendency for some individuals to put on weight, as they get older. Many adults also appear to age prematurely: greying hair and a gaunt facial appearance are common even in the early to mid-twenties. Medical characteristics specifically associated with WS include the following:

**(i) Cardiac Problems:** Cardiovascular abnormalities characteristic of WS include narrowing of the walls of the main blood vessel carrying blood from the heart (supravalvular aortic stenosis), and peripheral pulmonary artery stenosis. In a recent study of adults about two thirds were reported to have some form of heart problem. The severity of the cardiac anomalies varies from individual to individual, although it seems that more serious problems, requiring regular medication or major surgery, affect only a small minority (around 4%). However, it is strongly recommended that GPs are made aware of any history of cardiac problems, and that all adults with WS receive regular and routine check-ups. Carers frequently report that affected individuals have less physical vigour and tire more easily as they get older. This may be associated with the progression of congenital heart defects.

**(ii) Hypertension:** High blood pressure is frequently reported in the medical literature on WS, although it may go undetected in affected individuals. Again it is recommended that regular and routine checks are carried out by GPs, since hypertension increases the likelihood of a stroke and of other heart problems as individuals get older. Therefore, early diagnosis and treatment are important should any problems occur.

**(iii) Renal Problems:** Renal artery stenosis and renal hypertension are occasionally associated with WS; this should be borne in mind in cases where affected individuals are found to have hypertension. Investigation of the renal tract, for example by renal ultrasound, is advisable given the possibility of renal anomalies.

Urinary tract infections and cystitis have been reported in quite a number of cases. Some adults with WS report bladder and urinary tract problems such as excessive urination and difficulty with bladder control which can be a significant problem on long journeys to and from school, college or day centre. Many individuals urinate frequently, and unless they are clearly trying to gain attention or avoid a task, their requests for the toilet are usually valid. A few individuals seem not to recognize the sensation of needing to go to the toilet until the last minute – or even until they begin to wet themselves (although the latter is quite rare). Difficulties with toileting during the day or at night (wetting or soiling) may persist from childhood or appear as a new problem in adulthood. These problems may be exacerbated by anxiety or excitement. Carers may wish to enlist specialist help from psychologists or other professionals in Adult Learning Difficulty Teams in order to introduce a systematic toileting programme to address these problems.

**(iv) Bowel and Gastrointestinal Problems:** Although constipation is a major problem in WS children, this tends to improve somewhat in adulthood. For example, in a recent study of adults just over half (54%) had some bowel problems although these were severe in only 5%. Prolapse of the rectum, diverticulitis (sometimes caused by poor diet), and haemorrhoids can all cause problems. Compared to early childhood, hernias are comparatively rare in adulthood.

**(v) Joint Problems:** Commonly found skeletal abnormalities include limited rotation of the forearms (radioulnar synostosis), curvature of the spine (scoliosis) and contractures. In some cases these may progressively interfere with daily activities over time, and curvature of the spine may become more prominent with age. Many adults with WS complain of aches in the limbs, and joint dislocation (particularly in the knees) sometimes occurs.

**(vi) Menstrual Problems:** Early onset of menstruation is well documented in females with WS. As they get older, many women experience difficulties coping with the effects of the menstrual cycle. Premenstrual tension (PMT) is commonly reported, and individuals can become extremely irritable or tearful, sometimes with accompanying outbursts of anger. There is some evidence to suggest that periods may become more irregular with age, although premature onset of the menopause has not as yet been reported to be a significant feature.

**(vii) Hypercalcaemia:** A proportion of infants with WS are found to have high levels of calcium in the blood. These levels return to normal with dietary treatment, or simply with the passage of time. High calcium is found almost exclusively in infancy and regular checks of blood calcium later in life are unnecessary.

**(viii) Hypersensitivity to Sounds:** Hyperacusis refers to over-sensitivity to particular sounds that would not cause discomfort in most people. In a recent survey, we found that around 80% of adults with WS show at least some hypersensitivity to sounds. These seem to fall into two categories: sudden 'explosive' sounds (such as fire-works, balloons bursting, thunder, objects falling), and electrical noises (such as vacuum cleaners, drills, food processors, washing machines). In some cases even people's voices or laughter may cause individuals to become anxious or upset. The basis for hyperacusis is not clear. In many cases it appears to improve with age, but about 30% of adults continue to experience significant problems with hypersensitivity to noise. Individuals tend to cover their ears or try to avoid distressing sounds by leaving the room or switching off the radio or television. Panic attacks and aggressive outbursts have also been noted in some individuals, apparently in response to the discomfort caused by sounds. Many find noisy rooms or rowdy environments distressing, and in these cases it is advisable to reduce the level of noise in the work place or home setting as much as possible.

### **COPING WITH HYPERACUSIS:**

**The cause of hypersensitivity to noise is not fully understood, but there are certain things that can be done to make it less distressing:**

- 1. Reassurance and a clear and simple explanation about the source of the noise often help.**
- 2. The reactions will often diminish if individuals are able to exercise some control over the sounds that cause discomfort. For example, encourage them to use the vacuum cleaner, or to help with household chores by turning on the washing machine or food mixer.**
- 3. Individuals may be reassured if they know that they can leave the room for a few minutes at any point, if they are exposed to a distressing noise.**
- 4. Repeated gentle exposure to the sound may help the individual get used to it. For example, tape record one or more distressing sounds and encourage the individual to play back the tape, quietly at first, then gradually increasing the volume.**

Once individuals reach adulthood they no longer qualify for paediatric care and check-ups. Although some continue to see the paediatrician well into their twenties, most will depend largely on their GP for medical care in adult life. Since GPs are unlikely to have encountered anyone with WS before, it is extremely important that they are fully informed about the individual's medical history and about the medical problems generally associated with WS in adulthood. An Information and Health Surveillance pamphlet designed for GPs is available from the Williams Syndrome Foundation, which provides useful information about the potential health risks associated with the condition. It is recommended that renal function, cardiac function and blood pressure are checked routinely, at least once a year.

### **ABILITIES OF ADULTS WITH WILLIAMS SYNDROME**

People with WS have learning difficulties, although these vary in severity. About half have severe learning difficulties, while the remainder has mild or moderate learning difficulties. Most also show a distinctive pattern of abilities, often with relatively good spoken language and large vocabularies, but poorer perceptual and motor skills, and concentration difficulties.

### **ABILITIES OF ADULTS WITH WS:**

- i. Mild to moderate learning difficulties (although a small minority, around 3% have an IQ in the normal range).**
- ii. Relatively good spoken language but limited comprehension.**
- iii. Fluent and articulate speech, incessant chatter.**
- iv. Marked gross and fine motor problems.**
- v. Marked visuo-spatial difficulties.**

## **Language and Communication**

The language and communication skills of adults with WS are often relatively good, and quite out of keeping with their general level of learning disability. On the surface at least, most appear fluent and articulate, and have sophisticated and wide ranging vocabularies. They tend to use complex and grammatically correct sentences, incorporating long words, clichés and sophisticated phrases, which they have heard other people use. However, their speech can also be inappropriate and repetitive, especially when talking about topics of particular interest to themselves. In addition, they tend to exaggerate and over-dramatize events in conversation with others. For example they may describe minor mishaps as major tragedies.

In many cases their understanding of language is more limited than their expressive language would suggest, and inappropriate language may result from their use of words without fully understanding their meaning. For example, they may be able to recite instructions or directions word for word, (eg “you shouldn’t go off with strangers” or “look before you cross the road”) yet have difficulty understanding the most basic concepts (or putting their words into practice). In conversation, many adults lack the intellectual ability to imagine what other people may be thinking or to see somebody else’s point of view. Many find it difficult to build a conversation based on the dialogue of others; consequently their speech tends to be one-sided, which may well antagonize or irritate others.

People with WS will often use frequent chatter and incessant questions to initiate and maintain social contact with other people. They are very adept at using their superior verbal skills to get attention (occasionally this might include telling the most elaborate and untrue stories to elicit the attention or sympathy of others!). It is important to encourage individuals to answer questions relevantly and to engage in appropriate conversation with people they meet; it is also important to discourage them from chatting too much, asking irrelevant or inappropriate questions or engaging in conversation with strangers.

Many adults with WS talk to themselves at times, and this can occur in public as well as in private. This behaviour is more evident when an individual is anxious or annoyed, and can also serve as a distraction in stressful situations. It is unlikely to be a symptom of psychiatric disturbance, but should be discouraged.

### **DISCOURAGING EXCESSIVE CHATTER & INAPPROPRIATE SPEECH:**

**There are various ways of discouraging repetitive questions or excessive chatter about favourite topics or preoccupations:**

- 1. The adult may be told that his/her questions will be answered once and once only, and that further questions or comments on that topic will not be responded to.**
- 2. Conversations about special interests and preoccupations should be permitted only after a period of reciprocal conversation.**
- 3. Alternatively, specific times may be set aside for talking about special interests.**
- 4. Another useful strategy is to insist that individuals answer their own repetitive questions.**
- 5. Strategies for discouraging inappropriate speech will work only if the attention the individual previously received for inappropriate speech is replaced by attention for socially appropriate utterances. If this can be done consistently, inappropriate utterances will decrease in frequency.**
- 6. Parents and carers should point out to individuals when they are talking to themselves and ask them to stop, or distract them with some other activity. Another alternative is to have a very simple WRITTEN chart pinned up with the answers provided (in picture form if necessary) so that the person with WS can go and check for themselves.**

### **Perceptual And Motor Abilities**

Many adults with WS have particular difficulties with gross and fine motor coordination; with visual discrimination tasks, with orienting themselves and objects in space, and with judging distances, directions and depth. Even something as apparently simple as walking down stairs or on an uneven surface like grass or gravel, may prove extremely challenging. A fear of heights and unsteady surfaces is common, and crossing the road can be a particular problem, because individuals often find it difficult to judge accurately the speed and distance of cars. Difficulty with fine-motor coordination may make individuals appear clumsy or slow when performing delicate or intricate tasks, such as doing up buttons or threading a needle. Some individuals have a marked hand or arm tremor when performing such tasks. Handwriting is also affected by these difficulties.



**SUGGESTIONS FOR IMPROVING VISUO-SPATIAL & MOTOR SKILLS:**

- 1. Make use of relatively superior language ability to improve perceptual motor skills. It often helps if individuals are encouraged to talk themselves through each step of a task while they are doing it. This can help them to focus their attention on the task, and provides verbal reinforcement for activities.**
- 2. Incorporate objects and themes of particular interest into teaching materials. For example, encourage someone who is fascinated by cars or machines to copy and draw outlines of these to improve fine motor and perceptual skills.**
- 3. Try to incorporate music into appropriate gross and fine-motor exercises such as balancing and ball play. Dancing and music-led physical activities can help people better appreciate how their own bodies respond and behave in space.**
- 4. There are many enjoyable computer programmes available which help to develop matching and discrimination skills, and improve concentration and attention.**

**Concentration and Attention Span**

Poor concentration and over activity are common in children with WS. Over activity tends to diminish in adolescence, but distractibility and limited attention span can remain a problem, and adults with WS often find it difficult to concentrate on tasks for long periods. Their anxiety and hypersensitivity to noise can exacerbate this distractibility.

**ADVICE FOR BUILDING UP CONCENTRATION SPAN:**

- 1. Self-instruction can be a useful strategy. Individuals should be encouraged to remind themselves to keep working on the task at hand, initially by thinking out loud and later by silent self-instruction. So while carrying out a task the adult is taught to tell himself at regular intervals to "keep working" and to "concentrate on what I'm doing".**
- 2. Break down tasks into short, simple steps with cues - such as pictures or written instructions - to indicate clearly what needs to be completed at each stage.**
- 3. Give a reward for each step that is successfully completed.**

## **Reading**

The abilities of adults with WS vary considerably; about 50% do learn to read, at least at a basic level. Individuals tend to have relatively good verbal skills, including a very good memory for sounds and words, and excellent auditory sequencing skills. These strengths are useful for acquiring reading skills. Many individuals with WS reach their ceiling of reading ability in their mid-teens, and often do not progress beyond a reading age of 8 or 9 years; however this level provides a good basic reading vocabulary.

### **TIPS FOR IMPROVING READING SKILLS:**

- 1. Try and use existing skills in different contexts; for example, teach the individual to read timetables, shopping lists or menus, as well as newspapers and magazines.**
- 2. Encourage individuals to read magazines about topics or objects they are particularly interested in, such as cars or music.**

## **Writing and Spelling**

The majority of adults with WS attain at least some basic writing ability, such as writing simple sentences. However many find writing more difficult than reading because of the additional visual and fine motor skills required. Many people have problems holding a pen or pencil, and forming the letters, spacing out words, and aligning words. Again, only limited progress in spelling ability is found in the period from adolescence to adulthood.

### **TIPS FOR IMPROVING WRITING SKILLS:**

- 1. Increasing access to computers is extremely useful for encouraging adults who have particular difficulty with fine motor control to practice what would otherwise be a slow and laborious task.**
- 2. Many individuals have difficulty coping with the reading and writing components of a college course or work. Encourage them to dictate work to parents or tutors, or into a tape recorder, or introduce them to using a word processor as a way of bypassing the difficulties with writing.**

## **SOCIAL RELATIONSHIPS**

### **Over-friendliness**

Almost all adults with WS find it difficult to establish and maintain friendships with people of their own age, especially as they tend to be reluctant to socialize with other people with disabilities. On the other hand, they love company, are eager to please and will often seek out people and engage them in conversation. Unfortunately, more able adults of their own age may lack patience, and quickly tire of their repetitive chatter and limited conversation. Individuals with WS may approach strangers in an over-friendly and over-familiar manner, and will often tag along with them. This can be a major worry for parents and carers, who fear that they are too trusting and could be taken advantage of if not supervised sufficiently. Individuals with WS may occasionally seek affection and physical contact in ways that would be acceptable in a child but which are inappropriate in an adult. Again, such behaviours usually illustrate poor understanding of the boundaries of different types of relationships. Whereas hugging, kissing and touching are acceptable in children, this type of over-demonstrative behaviour may be a real barrier to acceptance in the ordinary community.

### **Understanding Other People**

As already noted, adults with WS often possess good verbal skills which enable them to initiate superficial social contacts. However, they often lack understanding of the underlying, 'unwritten' rules governing all types of social interaction. They are often too open, direct or personal in their communication with others, without recognizing the social constraints that would be apparent to other people in the same situation. In other cases their social naiveté and lack of inhibition can lead them to tell tales or to say things that might hurt or embarrass other people. Such behaviour is rarely intentional or malicious, but occurs because individuals do not understand the social implications of their utterances. Similarly they will not hesitate to try and gain other peoples' attention with their comments or questions, or to reprimand others. They may even try and imitate the manner of their supervisor when bossing or telling somebody off! Consequently they may give the impression of being rude, bossy or attention seeking, which again may antagonize others, if they are not forewarned.

Inability fully to understand the subtleties of social interactions, and a tendency to take language very literally, may result in individuals becoming easily offended or upset; even mild teasing or 'leg-pulling' can cause great distress. Many adults with WS believe, quite incorrectly, that they are being 'picked on', and are extremely sensitive to the perceived disapproval of others.

Concern for the physical and emotional well being of others can be one of the most endearing aspects of the personality of adults with WS. However, whereas they seem to know *when* people are upset they do not always understand *why* people feel that way, and some individuals are over-sensitive, believing that they have caused upset in others without understanding the true cause.

#### **COPING WITH OVER-FRIENDLINESS:**

- 1. From the earliest years, parents and carers should constantly reinforce the message that the individual should not approach strangers. When the individual does approach strangers, parents and carers should strongly indicate their displeasure.**
- 2. Whether meeting a familiar person, or when introduced to someone new, the individual with WS should be taught how to greet others in an appropriate way (shaking hands etc.). Over-enthusiastic and inappropriate behaviour, such as hugging and kissing strangers, should be firmly discouraged. It also helps if the recipient of the over-demonstrative behaviour is asked to make it clear that such behaviour is unacceptable.**
- 3. To encourage social skills, such as greeting people appropriately, social situations can be rehearsed in 'role-play' with individuals or in groups. Many adults with WS show an aptitude for drama, and this can be helpful in teaching them how their behaviour affects others and to provide appropriate 'models' of social behaviour.**
- 4. Prolonged and direct eye contact can be very disconcerting. Using video or role-play techniques, individuals can be taught simple rules, such as not to stand too close to others and not to stare.**
- 5. Remember to give plenty of gentle feedback and to praise adults when they behave in a socially appropriate manner.**
- 6. Do not encourage behaviours that may have seemed 'cute' or 'endearing' in childhood (such as hugging or kissing others) but in adulthood can lead to rejection, exploitation or even abuse. It is important to be firm with the individual, and to point out that such behaviours are unacceptable.**

### **DEALING WITH SOCIAL PROBLEMS**

- 1. If it appears that an individual does not fully understand the implications of a social situation, explanation and reassurance can often help to shape more appropriate behaviour.**
- 2. Carers and supervisors should simplify their conversation and instructions in order to ensure that individuals fully understand what is being said. Very often individuals behave in a socially inappropriate way because they do not understand or are confused about what is required in specific situations.**

### **Emotional Attachments**

Many individuals with WS do have boyfriends or girlfriends. Opportunities to meet members of the opposite sex may come at college, work or in social situations (e.g. Gateway Club). As these situations are often supervised, the resulting relationships are frequently rather 'superficial', although they may bring considerable satisfaction to the individuals concerned. Occasionally adults with WS may focus their affection on 'pop-stars', television/film personalities or someone they know like a neighbour or carer; where such emotional attachments become intense and obsessive, they can be extremely disruptive.

### **HOW TO DEAL WITH INFATUATIONS:**

- 1. Carers should gradually reduce the amount of time the adult spends talking about or having contact with the subject of the infatuation. Allowing limited contact at set times should help to reduce any anxiety that such restrictions may cause.**
- 2. The person who is the object of the infatuation should not encourage it in any way. Very often a sympathetic or understanding approach only serves to encourage unwanted attention. Individuals should be told firmly, and in terms they can understand, what constitutes acceptable behaviour and what does not.**

## **Sexual Relationships**

As is the case for many people with learning difficulties, adults with WS are often socially vulnerable, and their friendly and trusting nature may place them at particular risk of exploitation. Sex education at an appropriate level for the individual concerned, is crucial from an early age.

Some adults may display inappropriate sexual behaviours such as masturbating in public. The best way to deal with such behaviour is to teach the individual to identify when and where masturbation is appropriate (e.g. in the bedroom with the door closed) and where it is not (e.g. in public places or in the presence of others), rather than trying to stop it altogether. Teach that masturbating is not forbidden altogether, but is acceptable in private and at certain times.

The Foundation for People with Learning Disabilities ([www.learningdisabilities.org.uk](http://www.learningdisabilities.org.uk)) publishes some useful leaflets/booklets on sexual and relationship problems.

The subject of contraception raises a number of problems for adults with WS. Mechanical barriers are often impractical and ineffective. Given the risks associated with congenital heart disease, which is present in many cases, the advice of a cardiologist should be sought with respect to other forms of contraception. Where the oral contraceptive is contemplated for a woman with WS a low dose oestrogen should be used. Other alternatives include contraceptives given by injection or in capsules, which are slow releasing and long lasting.

Seeing friends and siblings leading independent lives, having relationships and getting married can be a source of frustration for adults with WS, who often cannot understand why they have been left behind. The desire to get married and lead an 'ordinary life' can lead some individuals to get involved in relationships, and even get engaged, without having a true understanding of what this involves. It is important that carers try to teach individuals about the subtleties of relationships with others, and help them to cope with the emotional difficulties that often occur.

## **EMOTIONAL AND BEHAVIOURAL DIFFICULTIES**

Emotional and/or behavioural difficulties are commonly reported in individuals with Williams syndrome. These include a wide range of issues, such as preoccupations and obsessions, impulsivity, distractibility, hyperactivity, eating and sleeping difficulties and attention seeking behaviours. Perhaps most notable, however, are problems relating to anxiety. Research has indicated that individuals with Williams syndrome show high levels of anxiety (with rates as high as 96% being reported) that occur throughout life.

### **Anxiety**

Adults with WS are often described as being over-anxious and easily upset by criticism or frustration, or by events that other people regard as trivial. They may worry excessively about the well-being of others, their own health or about all kinds of imagined disasters. In some cases this might manifest itself as hypochondria and exaggerating everyday aches and pains into serious illness.

Anxiety may result from many different situations, but three areas in particular tend to give rise to difficulties:

**(i) Excessive demands:** The typically good verbal abilities of adults with WS can lead others to over-estimate their general ability, and this can result in them being exposed to situations in which they are unable to cope. These can include living arrangements without adequate support, or college/work centre activities, which are too demanding. Inappropriate demands can cause individuals to worry excessively and to become anxious or upset, if only because they think that they might be upsetting or disappointing their parents, carers or supervisors. There are a number of indicators that suggest when someone is not coping: they may try to get out of doing things by complaining of headaches or tiredness; they may 'escape' stressful situations by having tantrums or being aggressive; they may repeatedly ask for reassurance from others, or simply walk out of the room. If the excessive demands persist, adults with WS may withdraw into themselves, lose interest in food, activities and/or relationships, and become depressed.

**(ii) Change and Uncertainty:** Individuals can become anxious when anticipating something new or different from their normal routine (even a visit to the doctor). Changes of staff or in the tasks undertaken at work can be particularly disturbing.

**(iii) Threat:** Many individuals with WS have difficulty understanding the complexities of social interactions, and although increasing integration into mainstream society is generally a very positive trend, it is important to be aware that the outside world can also be a hostile and frightening place. Adults with WS can become extremely anxious if they feel that they are being 'picked on' or if someone is behaving in a confusing or threatening way. They can also be very protective of the people they care for (family, supervisors, carers) and may become anxious if they think that someone or something is threatening them in some way.

Anxiety can show itself in many different ways: moodiness or apathy; tantrums; eating difficulties (e.g. excessive or fastidious eating or going off food); sleeping difficulties (e.g. restlessness or nightmares); over-sensitivity to criticism or disapproval; sudden loss of continence; picking and scratching the skin (sometimes causing bleeding) and increased stereotyped movements (e.g. rocking, thigh rubbing). Of course, individuals may get anxious for many other reasons than those described above, and anxieties may manifest themselves in many different ways.

Occasionally anticipatory anxiety can develop into a phobia -a persistent and intense fear of specific objects, situations or events, leading to avoidance of the feared object or situation. Phobias can develop from anxieties about noises (e.g. fear of balloons, dentist's drill or thunder) or upsetting experiences (e.g. phobias of needles dating from childhood visits to hospital).

**COPING WITH ANXIETY:**

- 1. It is advisable for parents and carers to strike a balance between comforting and reassuring the individual without making too much fuss (which may easily exacerbate feelings of upset and worry). Spend just a few minutes reassuring the individual and then move on to another topic of conversation or task.**
- 2. Anticipatory anxiety can often be reduced by spending a brief period ahead of time explaining and talking through what the individual is anticipating, and discussing the reasons for and the possible (realistic) outcomes of change.**
- 3. If an individual has a phobia, which is causing significant disruption to daily life, advice and help can be obtained from your local community mental health team, which works with adults with Learning Disabilities or the local Clinical Psychology Service. These services can be contacted directly or through the GP.**

In addition to anxiety, issues in other areas are beginning to become apparent. In a recent survey of adults with WS and their parents, we looked at a wide range of emotional/behavioural problems and found that 77% of parents reported that their son/daughter was experiencing some form of emotional/behavioural problem that was causing distress or intrusion into everyday life. These issues included fears and phobias (48%), anxiety (47%), obsessions and compulsion (30%), eating difficulties (19%), mood disturbance and depression (10-17%), hypochondriasis (10%), hallucinations (10%) and excessive emotion (5%). In follow up interviews with both parents and the WS individuals (using an assessment measure designed specifically for adults who have learning disabilities), we found that 29% of the adults with WS received a diagnosis of at least one mental health problem, such as specific phobia, depression, agoraphobia, schizophrenia, panic disorder, social phobia, psychosis and generalized anxiety disorder. Further, 60% of people who received a diagnosis met the criteria for at least one other psychiatric disorder.



## **Preoccupations**

Adults with WS often display intense fascinations and preoccupations with certain objects (e.g. cars, electrical appliances, animals, tools and machinery), particular topics (e.g. disasters and violence in the news, crime, their own health and the health of others, sports, future events such as birthdays or holidays) or an intense interest in particular people (pop-stars, television personalities, film stars, or people they know -such as a particular neighbour, carer or supervisor). They may spend a great deal of time absorbed with these topics and talk about them repetitively. Such preoccupations can bore or irritate other people and often serve to isolate the individual.

### **HOW TO DEAL WITH PREOCCUPATIONS:**

- 1. Try to keep the preoccupation within acceptable boundaries. Attempts to stop these interests completely are likely to be counter-productive, because the individual may become very anxious and distressed.**
- 2. Limit the amount of time spent in obsessional activities (or talking about them). Confine it to a fixed time (say 10 minutes a day).**
- 3. If the individual is not able to control the preoccupation, some of this special time should be forfeited.**
- 4. Once the preoccupation is under better control, try to ensure that the time that used to be spent with obsessions is filled with other, more appropriate activities.**

## **Repetitive Movements (Stereotypies)**

Some individuals with WS display persistent, repetitive movements (stereotypies) such as rocking or hand flapping. These behaviours are often found in people with learning difficulties, and may be particularly prominent when the individual is tired, anxious or agitated. In some cases rocking may occur when someone is engrossed in a particular activity, and it may aid concentration. However such behaviours can also limit social acceptance. Generally it is advisable to make individuals aware of their rocking or hand flapping as soon as it begins, and to ask them to stop.

**DEALING WITH STEREOTYPED MOVEMENTS:**

- 1. At first the individual will need plenty of reminders to stop the behaviour, although with time a nudge or pre-agreed word will usually suffice.**
- 2. It is often helpful to combine this approach with a reward system whereby individuals are rewarded if they can spend increasing periods of time without displaying any stereotyped behaviours.**
- 3. Encouraging them to change seating position or the type of chair they usually sit on may also be effective.**
- 4. Distracting individuals can help to reduce stereotyped movements when they occur.**

**Moodiness and Aggressive Behaviour**

Frustration or being unable to get one's own way can sometimes lead to excessive displays of annoyance, outbursts of anger and, at times, aggressive behaviour such as hitting out. Such displays can occur at home or in public places, and can represent a major management problem for parents and carers.

### **ADVICE FOR DEALING WITH AGGRESSION OR ANGER:**

- 1. Firm and consistent handling is important. Carers and supervisors should not give in to the adult's demands, and should not reward such behaviour with attention or access to desired objects or activities. Otherwise individuals will quickly learn that having tantrums is an effective way of getting what they want.**
- 2. An effective approach is to remove individuals from the setting where the tantrum took place, and make it clear that they cannot return until they have calmed down. As soon as they are calm, praise them for doing so and allow them to rejoin everyone else.**
- 3. Observe the individual carefully when tantrums or outbursts occur and try to identify the events and situations that trigger such behaviour. If a particular trigger can be identified this can either be avoided or the individual's attention can be distracted onto something else before the tantrum has a chance to develop.**
- 4. Try to adopt exactly the same approach each time a tantrum occurs, and try and ensure that others adopt the same approach, as you - a consistent response from everyone is crucial.**
- 5. Reward appropriate behaviours. If you cannot allow someone to get their own way, take a couple of minutes to explain why. It may take a little while for the individual to learn the new rules about tantrums, but a consistent approach can be remarkably effective.**
- 6. Aggressive behaviours can become more complex, and it may become difficult to identify triggers. In such cases advice should be sought from local Psychology Departments and Adult Learning Difficulty Teams, which can be contacted directly or through the GP.**

## **INDEPENDENCE AND SELF CARE SKILLS**

The development of independence skills becomes increasingly important in adulthood -both for the WS individuals and for their families. Parents, carers and supervisors should provide opportunities for individuals to maximize their potential, but in a safe environment, which does not place excessive demands on them. The particular characteristics associated with WS can both limit and facilitate independence. For instance, an adult may be able to communicate with others, which obviously facilitate independence, but still display an over-friendly or anxious personality, both of which limit the amount of social independence that can be safely allowed. Such patterns of relative strengths and weaknesses are characteristic of individuals with WS, and can make it difficult for parents and professionals to judge how much independence is suitable in each case.

### **Living Arrangements**

In a recent survey of adults with WS aged 18 to over 50 years, we found around half still lived with their parents, and 40% lived in sheltered accommodation, group homes, residential communities) or residential colleges. Very few lived independently or semi-independently. These figures indicate that most adults with WS continue to need at least some supervision and support in their daily lives. Even the people who lived independently needed regular help from relatives to cope with housework, budgeting and self care.

Although the choice of living arrangements is still somewhat limited, there are a number of charitable and other organisations, which offer a range of different provision. The Learning Disabilities Team at local Social Services Departments will provide information about sheltered and community based accommodation available locally. Regional MENCAP organizations are another extremely useful source of advice.

Moving away from home can be a difficult as well as an exciting time for young adults with WS. Because of their over-anxious personalities and dependence on their families, they may find a change of home environment particularly worrying, and even traumatic. This does not mean that such a move should be avoided, but rather that it should be carefully planned, with support from the family to help the individual cope in the new environment. Many parents believe that it is better to help their son or daughter settles into alternative long term accommodation while they are still fit and well, rather than waiting until they are no longer able to care for them.

Anthony Quinn's "Guide to Families Wishing to Make Legal Provision for a Learning Disabled Member" (see below) provides helpful advice on planning for the future financial and legal welfare of a learning disabled son or daughter.

### **Respite Care**

Respite care is short-term residential care provided by local Social Services where individuals with special needs can spend a few weeks or days in a special unit or with another family. Respite care is helpful not only for giving parents and other carers well-deserved breaks at regular intervals, but also for enabling the individual to experience different environments, meet different people and gain independence skills. The standard and availability of respite care services is variable nationwide. Parents/carers can obtain information on what is available locally from social workers (Learning Disabilities Social Services), from special needs schools and colleges, and from other parents of individuals with disabilities. The National Association of Family Based Respite Care (Norah Fry Research Centre, University of Bristol, 32 Tyndalls Park Road, Bristol BS8 1PY) publishes an annual survey of family-based respite care schemes available in the UK.

## Self Care Skills

As individuals get older they are expected to learn a whole array of skills while moving towards independence: washing, toileting and hygiene, bathing, shaving, dealing with menstruation, dressing, cooking and kitchen skills, undertaking household chores, shopping and dealing with money, telephone skills, and travelling. Many adults with WS require help in developing these skills. They may have problems in physically carrying out the required task or in fully understanding and planning the task requirements. They may also need help in deciding when or how often a task should be performed -for example, should one have a bath or change of clothes every day?

### **TIPS FOR TEACHING INDEPENDENCE IN SELF CARE:**

- 1. Taking a 'whole job' perspective helps people to get a better idea of what is involved. For instance teaching someone to bath includes running the bath, washing thoroughly all over, then letting the water out, and drying. Break the task down into small steps and teach one step at a time. For example, teach how to put on pants, then socks, then shoes, then tying shoelaces, concentrating on one step at a time.**
- 2. Use wall charts, a series of pictures or lists to clearly indicate the various stages of the task and the order in which they should be completed. A list of steps that have to be carried out for the completion of a task could be written out and rewarded each time they are completed appropriately. Start off with just a couple of simple steps and when they are routinely established, gradually build up the sequence.**
- 3. Establish routines, and give rewards (such as stars or money tokens), to reinforce the idea that self care and hygiene must be practised regularly.**
- 4. Try and use verbal cues that can be repeated out loud by the individual. For example, when tying shoelaces teach the individual to say out loud "cross the laces over, pull one through, pull tight" to establish a routine.**
- 5. Music and tunes, for which WS adults appear to show a natural propensity, can also be used for establishing sequences and routines. Incorporating a sequence such as "every day after dinner wash your hands" -into a tune may help the individual remember it.**

## Community Independence

Adults with WS may be capable of learning to travel independently along familiar routes, or even further a field. Many parents and carers are reluctant to let individuals out alone because they are understandably worried about the possibility that the individual will wander off or approach

strangers. This can be a real danger, and it is therefore important that parents and other carers start as early as possible, to teach independent travel and safety when out.

**TIPS FOR ENCOURAGING TRAVEL AND INDEPENDENCE:**

- 1. Using the adults' good verbal skills, help them to memorize travel instructions and travel routes by talking them through. If someone has to learn a new bus route to work or college, it is a good idea to have a supervised trial run first before the individual tries it alone.**
- 2. If the adult can read, short written instructions will help.**
- 3. Wearing some sort of identification, such as an identity bracelet, can be helpful in the event of difficulties or an emergency. Medic Alert (Free phone: 0800-581420) is a charity which provides a bracelet or necklace engraved with medical details of the individual who wears it, a personal identification number and a 24-hour emergency telephone number.**

**Numeracy and Coping with Money**

Many adults with WS find dealing with numbers particularly difficult, and this can make coping with money a particular obstacle. Individuals with WS often have a poor concept of the value of money, and budgeting skills are usually extremely limited. Any ideas of saving can easily be swamped by their interest in particular items. Likewise, many individuals are attracted by mechanical or electrical equipment, and the 'charm' of fruit machines can often be an expensive distraction.

The caring nature of many WS adults and their naiveté can also make them vulnerable to financial exploitation. Because of this, most parents and carers tend to take charge of finances (including benefit payments), while giving the individual a daily allowance (weekly allowances are often 'frittered away' on the first day).

#### **TIPS FOR TEACHING MONEY SKILLS:**

- 1. Use real money (rather than tokens or pen and paper examples) in real-life situations. For instance, take the individual to the supermarket to buy food, or to a cafe to buy a cup of coffee.**
- 2. Encourage independence by gradually reducing the amount of supervision provided. For example, if the individual has the basic skills required to make small purchases, the next stage would be to stand at the back of the shop while s/he pays for the items. The next stage would be to stand outside while s/he completes the purchase.**
- 3. Discuss problems and use role-play and games to increase awareness of money.**
- 4. Encourage the adult to keep records of income and expenditure, as a means of gaining more control over budgeting.**

#### **Coping with Time**

Problems with numbers and visuo-spatial difficulties also lead to problems understanding time. For people who have difficulty telling the time using analogue clocks, digital displays may be easier. It is also helpful to link the time on the clock to activities that take place at specific times. Many individuals also have problems with the concept of time itself, and find it extremely difficult to judge approximately how long a minute, hour or afternoon is. Often they will use events, such as lunch or home time, to organize their day, and they can become extremely anxious if things do not happen exactly as they are supposed to.

They can also become upset for other apparently 'trivial' reasons, such as a bus being delayed or if they think they are late for an appointment. There may be particular difficulty with the concept of future time, and inability to plan or schedule their activities is a characteristic problem.

#### **Leisure Activities**

Adults with WS are naturally sociable and can gain a considerable amount of enjoyment and useful experience from leisure pursuits. However they typically need help to organize their leisure activities, because they tend not to show initiative in choosing or persisting with activities themselves. Many carers find that if someone else does not organize activities or outings, the individual will simply stay at home and engage in passive activities such as watching television or listening to music. If encouraged, however, adults do enjoy attending clubs designed for individuals with learning difficulties (e.g. Gateway or PHAB clubs), and some also belong to special interest clubs (e.g. train spotting or car clubs), which are related to their specific interests or preoccupations. In this way they can be introduced to new environments and social groups.

Many individuals with WS have considerable aptitude for music, and some play instruments, such as percussion and keyboards. Even those who cannot actually play, are often interested in music, and may have a remarkable propensity for recognition of tunes and accompanying words. These abilities too can be harnessed in order to encourage leisure and club activities. Adults with WS are generally not very good at gross motor activities like ball games and other sports. However many enjoy swimming, which is a good way to encourage physical exercise. The lack of interest in sport may be attributed to the complexities of social interaction involved in-group sports or to the increasing tiredness and lack of stamina reported in WS adults, as they get older. A number of parents report that lack of exercise is a contributory factor to the weight gain observed in many individuals in adulthood.

Information about local social and sports clubs for adults with a range of disabilities can be obtained from local colleges, adult education centres or from a social worker. Clubs often arrange supervised outings and other social activities, where people can meet together on a regular basis. A number of organizations (including the Williams Syndrome Foundation) arrange supervised holidays for disabled adults, and financial assistance for such holidays is often available.

## **LEAVING SCHOOL**

### **Further Education**

The question of what will happen after leaving school can be a major worry for families. Information about the various options available should be obtained well before school leaving age. Many Further Education and Technical Colleges integrate special needs courses into their prospectuses. Although the majority of families are satisfied with these further education courses, the quality can vary greatly from college to college. It is strongly recommended that parents find out as much as possible about the specific courses and colleges they are considering. Talking to the staff and the parents of students already undertaking particular courses is a good way to obtain information. Teachers in special schools are usually familiar with the quality of further education available locally, and school careers officers and social workers can also provide useful information.

Residential colleges for young adults with special needs are another option. These are extremely useful for providing an introduction to living away from home, and they may also provide work placements in the local community. However Local Authorities are sometimes unwilling to fund a placement outside of their own region unless it can be proved that an 'out of borough' placement is more relevant to the needs of the adult than anything available locally. Careers Guidance Services will provide information about training centres and residential colleges locally and further a field. They should have a copy of a publication called 'COPE' published by Lifetime Careers (address at the back of this booklet), which provides a comprehensive list of residential colleges and training centres catering for adults with learning and physical disabilities in Great Britain.

It is generally advisable for further education courses to concentrate on helping adults with WS to acquire and improve their daily living skills and to concentrate on pre-vocational training, rather than spending a lot of time on academic skills.

Young adults with WS may encounter a number of problems coping with further education courses. These include inability to cope with the demands of the course, and related problems such as punctuality, travelling and interpersonal difficulties. It is important that tutors and key-workers appreciate the pattern of strengths and deficits that characterize individuals with WS, and are aware of their particular behavioural and cognitive difficulties which may require particular support from staff (see section on Work-related Difficulties).



## **Jobs and Sheltered Placements**

Most adults with WS attend Adult Training & Education Centres or Day Centres, or remain at home with no daytime occupation. Only a minority is in paid work, and most of these are supervised work placements. When making decisions about training and occupations for individuals with WS, their profile of strengths and deficits needs to be fully understood. Jobs that are typically considered suitable for people with learning difficulties include routine manual tasks, such as stacking shelves, packing, or assembly line work. However, difficulties with visuo-spatial and motor skills, limited concentration span, and lack of physical stamina means that manual jobs such as these are often not suitable for people with WS. Individuals with WS also have marked difficulties coping with numbers, and jobs involving tills and dealing with money are often inappropriate. Adults with WS are usually extremely caring, and this concern for the physical and emotional well being of others may be utilized in employment in homes for the elderly or disabled, with appropriate supervision. Helping jobs (e.g. being a 'right-hand man') or service jobs (e.g. dealing with and helping the public in coffee bars or libraries) are other possibilities. Jobs that relate directly to the individual's special interests or preoccupations should also be considered. For example if the individual is very interested in cars then s/he may prefer to work in a garage, even if the actual job is in itself fairly routine.

Parents and carers may have to do a considerable amount of 'home-work' in order to obtain adequate advice about employment. Information about what is available locally can be obtained from a Disability Employment Adviser (DEA), who can be contacted through larger job centres. The DEA can also help make contact with national organizations such as MENCAP Pathway Employment Service or the Shaw Trust (national addresses at the back of this booklet) -both of which strive to provide employment opportunities for adults with learning difficulties. DEAs are part of the Department of Employment's Placing, Assessment and Counselling Teams (PACT), which can also provide advice about local schemes and organizations. Local Learning Disabilities Social Services Departments are also useful sources of advice about employment opportunities.

## **Work Related Difficulties and Suggestions for Suitable Work Placements**

In a recent survey of adults with WS, we found that just over a third were currently involved in some kind of work experience. These placements included sheltered jobs, voluntary work and job placements organized by Further Education Colleges or Adult Training Centres. Fewer than 10% were in independent regular paid work. About a third of the people in work placements were considered by their employers to have some problems in their job, but only a small minority was considered to have major problems. About one third of those in work placements required 'some extra supervision' in the workplace, one-third required 'considerable extra supervision' and the remainder required 'constant supervision'. The problems typically encountered in the workplace are very similar to those faced in the home and in social settings.

**(i) *Appropriate Demands in the Workplace:*** The superior speech and language abilities of adults with WS often disguise their much more limited level of intellectual functioning. Individuals often give the impression that they are more able than they actually are, and as a result, they may be placed in education and employment settings that place excessive demands on them. Equally, many individuals with WS may have little insight into their own limitations, and will often claim that they can carry out tasks far beyond their capability.

Supervisors report that many adults with WS can only cope with simple instructions in the workplace, and some need help to understand even at the simplest level. Very few are capable of understanding 'complex tasks'. Moreover, about half dislike changes in routine sufficiently to

cause at least some disruption to their jobs. Many individuals with WS lack strength and tend to tire more easily as they get older. Activities that are physically demanding or which involve a lot of standing can be extremely tiring. If an individual is complaining of fatigue or is obviously looking tired as the day or week wears on, it may be a good idea either to lessen the physical workload or reduce the hours worked. Our recent study of adults found that jobs were typically in shops, domestic or catering work (often in homes for the elderly or people with learning disabilities). Many families felt their son or daughter could have coped with a higher level (and better paid job) had better support and opportunities been available.

**(ii) Adequate Supervision:** Many adults with WS are extremely distractible and restless, and as a result require a high level of supervision in order to concentrate on a task. Many display intense preoccupations with particular subjects or people, which can also serve to distract them from their work. Supervision is needed to ensure that tasks are completed. Without adequate supervision, people tend to give up work they are struggling with, without asking for assistance. Some become disruptive if they are unable to cope. Unless carefully supervised, work tends to be 'slap-dash', and poorly organized.

Supervisors need to provide reassurance, simple explanations and demonstrations of what is required, prompts and reminders and help when trying new tasks or adapting old ones. Supervisors should also be prepared to give people with WS emotional support and reassurance should they become anxious or upset for any reason. Often adults with WS will only take instruction from someone they recognise as a supervisor. They tend to resent being told what to do by other people, which can cause problems with workmates. Having an approachable supervisor as a source of advice, reassurance and instruction can circumvent many of these difficulties.

Rewards can be given for completing tasks appropriately. Similarly, developing self-instruction is often a useful strategy for improving concentration. The individual should be reminded to keep working on the task at hand, initially by thinking aloud and later by silent instruction to him/herself. So while carrying out a task, the adult is taught to tell himself at regular intervals to "keep working" and to "concentrate on what I'm doing". Breaking down tasks into short, simple steps with cues - such as pictures or written instructions - to indicate what needs to be completed at each stage can also be useful.

**(iii) Reducing Anxiety:** Adults with WS tend to be particularly sensitive to the perceived criticism of others, and anxiety is a prominent personality characteristic. In the workplace, anxiety may result from excessive or unrealistic demands, or from uncertainty about the nature of the work. Individuals find it particularly difficult to adjust to changes in the way work is undertaken, as well as changes in personnel. Noisy and busy work environments may also result in increased distress due to their hypersensitivity to sound. Anxiety may manifest itself in the individual seeking constant reassurance from work mates and supervisors, increased stereotyped movements (eg. rocking, thigh rubbing, and hand rubbing), withdrawn behaviour or irritability and moodiness.

Exposure to situations that are stressful or over which individuals have little or no control can lead to tantrums and aggressive outbursts. Fortunately displays of anger and violent outbursts are rare, but where they do occur frequently advice from a psychologist working with people with learning difficulties may be helpful. If it is felt that the individual is more nervous or anxious than usual, it will be important to examine both work and non-work environments to ensure that the demands made are not excessive. Stress and anticipatory anxiety can often be reduced by spending a short period ahead of time preparing the adult for the task or feared event, and by explaining in detail what can be expected and talking through possible outcomes.

**(iv) Friendly and Sociable Environment:** Poor understanding of the 'unwritten' rules that govern social intercourse, of the boundaries of different types of relationships, and their keenness to please others, often result in adults with WS appearing to be over-friendly, inappropriately demonstrative and excessively chatty. Such behaviours can provoke irritation and even anger in colleagues and supervisors. Nevertheless most are accepted and well liked by their colleagues and supervisors.

Because many adults with WS find noisy or busy environments distressing, a quiet work environment, which is as free from distractions as possible, is recommended.

Individuals with WS often get particular pleasure from meeting and helping people, and often enjoy tasks such as running errands for others. Such tasks can be extremely rewarding and will also reduce boredom and distractibility.

On the whole adults with WS do not work well in teams, and often resent receiving instructions or directions from others. It is generally best to give individuals a particular area of work or lists of simple tasks that are their sole responsibility. This can also enhance feelings of self-esteem.

**(v) Structured (but not repetitive) Work:** Because of their distractibility and short attention spans, adults tend to get bored with repetitive work. Conversely they dislike change, and seem to work better when given a structured routine. More complex tasks can be coped with if individuals are provided with a written list or a sequence of pictures outlining task requirements.

### **Alternatives to Employment**

Paid employment is difficult to find for any one with a learning disability and may be beyond the capabilities of many adults with WS. Alternative daytime occupations include Day Centres and Adult Education & Training Centres for adults with special needs, run by Local Authorities and Social Services. Adult Training Centres tend to offer more structured activities, often including pre-vocational training, whereas Day Centres offer more recreational activities. As with most services, the quality of these centres varies enormously, and parents and carers are advised to find out about the quality of the provision available locally. Information can be obtained from local Learning Disability Social Services Departments.

### **IMPLICATIONS FOR SIBLINGS OF ADULTS WITH WILLIAMS SYNDROME**

Living with a brother or sister who has WS can be rewarding and fun, but also confusing and stressful. The needs of individuals with WS can at times be overwhelming, and they often require a disproportionate amount of the family's time and attention. As a result, brothers and sisters may get less attention from parents, grandparents and other relatives, and they may quite understandably feel resentful or angry. Parents need to try and ensure that they give their other children as much individual time and attention as possible.

Siblings may feel embarrassed or resentful when, for example, they have to explain to their friends about their disabled brother or sister, or when having to keep an eye on him or her. At times they may even feel guilty that they themselves are to blame in some way. Typically they also feel a great sense of responsibility towards their brother or sister who has WS, and as they

become adults they may worry about the future and who will look after their sibling once their parents are no longer able to do so. Realistic planning for the future can help alleviate many of these justifiable concerns.

Many siblings are poorly informed about WS, and as a result may harbour unnecessary worries about the possibility that they may be at risk of having a child with WS themselves. It is important for parents to talk with their unaffected children about WS, and to give them information about the cause of the condition, the negligible risk of recurrence in families, and to explain the physical and behavioural characteristics associate with WS. Siblings may also find it helpful to talk with the WS child or adult's paediatrician, cardiologist or clinical geneticist. Siblings have as much need for information as do parents, and accurate information will serve to allay many of their worries and fears.

Throughout this booklet we have emphasized the importance of independence in adulthood, and of providing a safe yet stimulating environment to enable people with WS to make the most of their potential. Getting the balance right between hanging on and letting go can be difficult. Close cooperation between parents and professionals who will help provide living arrangements, daytime occupations and leisure activities can make the task much easier for the individuals and their families. Very few people will have encountered an individual with WS before, and parents and carers play a critical role in providing information and advice on the most appropriate ways of meeting the special needs of adults with WS.

## USEFUL ADDRESSES AND FURTHER READING:

### **General:**

#### **Foundation for People with Learning Disabilities:**

([www.learningdisabilities.org.uk](http://www.learningdisabilities.org.uk)) Publishes many leaflets, booklets and books on general issues related to learning disabilities as well as specific areas including education, employment, accommodation, health, friendships, sexual relationships, leisure, respite care, etc.

#### **Mencap (Royal Society for Mentally Handicapped Children and Adults)**

Mencap National Centre  
123 Golden Lane  
London EC1Y 0RT  
Tel: 020 7696 5554  
[www.mencap.org.uk](http://www.mencap.org.uk)

Provides information about residential homes and other accommodation for adults with disabilities. Also provides details about the national network of Gateway clubs, Pathway Employment Schemes, and legal, benefits and welfare advice.

**Adult Learning Difficulties Teams** - available in most NHS Trusts or NHS Community Units. They tend to be multi-disciplinary teams consisting of psychologists, occupational therapists, speech therapists, psychiatrists and community workers. In some cases, social workers are also members of such teams. These professionals can give valuable advice concerning abilities and behavioural and emotional difficulties of adults with WS.

### **Further Education Colleges and Training Centres:**

**COPE:** Director of post-16 residential education and training for young people with special needs. 2006.  
Can be ordered through **Mencap** (address above)

Published by:  
**Lifetime Careers, Wiltshire Ltd**  
7 Ascot Court  
Whitehorse Business Park  
Trowbridge  
Wiltshire  
BA14 0XA  
Tel: 01255 716000

### **Assisted Employment:**

**Pathway Employment Scheme** (Mencap, address above). Regional network of Pathway Services around the country aiming to provide sheltered employment and supported work experience for adults with disabilities. Can be contacted locally through the Disability Employment Adviser in local Job Centres.

**The Shaw Trust:**

Fox Talbot House  
Greenways Business Park  
Malnesbury Road  
Chippenham, Wiltshire  
SN15 1BN  
Tel: 01225 716350  
[www.shaw-trust.org.uk](http://www.shaw-trust.org.uk)

Similar to Pathway, in that it aims to provide sheltered employment and supported work experience for adults with disabilities. Can be contacted locally through the Disability Employment Adviser in local Job Centres, or directly by phone.

***Residential Accommodation:***

**Mencap Homes Foundation** (Mencap address above)

**Home Farm Trust**

Merchants House  
Wapping Road  
Bristol  
BS1 4RV  
Tel: 01179 302600  
[www.hft.org.uk](http://www.hft.org.uk)

***Legal Provision:***

**Quinn, A.** (1997) "A Guide to Families Wishing to Make Legal Provision for a Learning Disabled Member" (2nd ed.) Available from The Smokery, Greenhill's Rents, London EC1M 6BN

***Personal Relationships and Sexuality:***

**Dixon, H.** (1988) Sexuality and Mental Handicap. An Educator's Resource Book. Wisbech, Cambs: LDA.

***Respite Care:***

**Family Based Respite Care**

Norah Fry Research Institute  
University of Bristol  
3 Priory Road  
Bristol  
BS8 1TX  
Tel: 01173 310987  
[www.bristol.ac.uk/norahfry](http://www.bristol.ac.uk/norahfry)

***Benefits and Entitlements:***

**RADAR (Royal Association for Disability and Rehabilitation)**

Unit 12 City Forum  
250 City Road  
London  
EC1V 8AF  
Tel: 020 7250 3222  
[www.radar.org.uk](http://www.radar.org.uk)

A campaigning and information-giving organization. Particularly useful for advice about benefits and entitlements.

**Disability Alliance**

Universal House  
88/94 Wentworth Street  
London E1 7SA  
Tel: 020 7247 8776  
[www.disabilityalliance.org](http://www.disabilityalliance.org)

**Family Fund**

Unit 4, Alpha Court  
Monks Cross Drive  
Huntington  
Yorkshire  
YO32 9WW  
Tel: 0845 1304 542  
[www.familyfund.org.uk](http://www.familyfund.org.uk)

Produce a booklet called "After Age Sixteen, What Next?" which gives advice about changes in DSS benefits and entitlements when individuals reach adulthood. The booklet is free for parents of individuals with special needs, though professionals are charged £4.00 to obtain a copy.

***Independence Training:***

**Carr, J. & Collins, J.** (1992) Working Towards Independence: A Practical Guide to Teaching People with Learning Disabilities. London: Jessica Kingsley Publishers.

**Yule, W and Carr, J** (1987) Behaviour Modification for People with Mental Handicaps (2nd edition). London: Croom Helm.

#### **FURTHER READING:**

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**Morris, C. A., Loker, J., Ensing, G., & Stock, A. D.** (1993). Supravalvular Aortic-Stenosis Cosegregates with A Familial 6 7 Translocation Which Disrupts the Elastin Gene. *American Journal of Medical Genetics*, 46, 737-744.

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## **ACKNOWLEDGEMENTS**

We would like to thank Sir George and Lady Cynthia Cooper and The Williams Syndrome Foundation for highlighting the need for an information booklet on adults with WS, and for funding the research upon which this booklet has been based. Alison Payne, Carol Mathurin, Cynthia and Ken Scott, Cynthia and Michael Glanville-Smith, Susan Golder, Christine Newman, Mike Adlam, Dr Mike Wolfman and Dr Neil Martin read and commented on earlier drafts of this booklet. Catherine Donnelly provided guidance concerning the vocational needs of adults with WS. We thank them all for their helpful suggestions. During the course of this research we have benefitted from insights and information provided by many parents, tutors, supervisors and key-workers of individuals with WS, and from meeting with and interviewing adults with WS themselves. We are grateful to them for all they have taught us about the condition.

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