

williams

**SYNDROME
FOUNDATION**

**WILLIAMS SYNDROME
GUIDELINES FOR PARENTS**

by

Orlee Udwin, William Yule & Patricia Howlin

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 a child with WS before, and they may understandably know little, if anything, about the characteristics and difficulties associated with the condition. The purpose of this booklet is to provide information about this rare disorder, and to outline the difficulties that may be experienced by children with WS. Of course every individual is different, and not everyone with WS will show all of the characteristics and difficulties described below. The contents of this booklet are based on research undertaken over the last twenty-five years, which demonstrates that WS is associated with a number of distinctive behavioural and psychological features, which differentiate children with this condition from others with learning difficulties. It is hoped that this information and the advice and suggestions that follow will be of benefit to the families of children with WS. A separate information booklet for teachers is available from the Williams Syndrome Foundation, and families may choose to forward that booklet to their child's school. Two additional booklets relating to adults "Guidelines for Families & Professionals" and "Guidelines for Employers & Supervisors" are also available from the Williams Syndrome Foundation.

Published by:
The Williams Syndrome foundation
161 High Street
Tonbridge, Kent TN9 1BX

Illustrations by Cynthia Cooper
First published in 1988.
Revised version published in 2007.
Copyright 1988, 1998, 2007.

Contents

Description of the Syndrome

Feeding and Eating difficulties

Sleeping Difficulties

Toilet Training

Dressing

Social Relationships

Anxiety

Preoccupations, Obsessions and Stereotyped Behaviours

Temper Tantrums and Aggressive Behaviours

Hypersensitivity to Sounds

Speech and Language Skills

Concentration Difficulties and Over activity

School -Related Concerns

1. Type of Schooling
2. Working with Teachers
3. Perceptual and Motor Difficulties
4. Reading
5. Leaving School

Implications for Siblings

Sources of Help

Acknowledgements

DESCRIPTION OF THE SYNDROME

Williams Syndrome (WS) is a genetically determined, intellectually disabling condition. Most affected children develop severe feeding difficulties in the first year of life, including vomiting, constipation and refusal to feed; they may be irritable, cry excessively and do not thrive well. Some of the children are found, on testing, to have raised levels of calcium in the blood (infantile hypercalcaemia); they will be put on a low-calcium and vitamin D-restricted diet by their doctor, and the feeding difficulties then improve, either rapidly or, in some cases, more gradually. Many of the children also have a distinctive facial appearance (the 'elfin' face) (see photographs), renal and cardiac problems (a heart murmur, narrowing of the walls of the main blood vessels carrying blood from the heart), which may be severe or quite mild. Other children, while having the typical facial appearance and heart problems, do not have high calcium, and so are not put on low-calcium diets.

Individuals with WS may also have dental abnormalities, back and joint problems, raised blood pressure and a delayed rate of growth, including low stature and a slight build. The average adult height is 5ft for females and 5ft 6ins for males.

Genetic characteristics

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. Deletion of this is associated with issues regarding connective tissue, such as heart disease, premature ageing of the skin and hoarse voice. 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.

Most cases occur sporadically, and the risk to parents of having another child with WS is no higher than the original risk. Brothers and sisters of WS individuals, too, are not at any increased risk of having children with WS. However, people with WS themselves have a 50% chance of transmitting the condition to their children.

Children with WS tend to be delayed in their development and show a range of learning difficulties, which in different children, can vary from mild to severe. In addition, they show a distinctive pattern of abilities and particular behavioural and personality characteristics which are common to most individuals with this syndrome, and which set them apart from other children with learning disabilities.

ABILITIES OF CHILDREN WITH WILLIAMS SYNDROME

- **Mild to moderate learning difficulties**
- **Relatively good verbal abilities**
- **Fluent and articulate speech by school age, often with incessant chatter**
- **Marked gross and fine motor problems**
- **Marked visuo-spatial difficulties**

The children tend to be very outgoing, sociable and affectionate, and by school age most develop fluent and articulate spoken language. They love talking and interacting with adults, and are helpful and eager to please. Many chatter incessantly, but at a superficial level, and often in a formal, adult-like way with frequent use of stereotyped phrases and clichés. Taking turns and keeping to the point in conversation may be especially difficult for them. In many cases their understanding of speech is not as good as their level of spoken language and this, together with their highly developed sociability may give the impression that the child is more able than is actually the case. In contrast to their relatively good speech, many of the children have visuo-spatial problems, and difficulties with gross and fine motor co-ordination. So they may be slower than usual in learning to sit and to walk, and they may find tasks like riding a bicycle, doing up buttons, cutting with scissors and holding a pencil particularly difficult to master. They tend to have poor posture, an awkward gait and limitations of joint movements. Because of their motor and perceptual problems they may also be fearful of heights and of negotiating stairs and nervous of uneven surfaces such as grass, gravel or sand.

Many children with WS are overactive and find it difficult to sit still and concentrate on particular tasks for any length of time. On the other hand, they can become fascinated and preoccupied by particular objects or topics and spend a great deal of time absorbed with these. While many are very friendly (and even over-friendly) to adults and seek out adult company, they may have difficulty making and keeping friends of their own age, often managing somehow to antagonise their peers.

BEHAVIOUR OF CHILDREN WITH WILLIAMS SYNDROME

Overactive
Limited concentration span
Sociable & outgoing
Overfriendly to adults

Excessively anxious
Preoccupied with objects/topics
Hypersensitive to sounds
Fearful of heights/uneven surfaces

Children with WS are often described as having anxious personalities and worrying excessively about themselves and others. Sleeping and toileting problems are fairly common. Many are also hypersensitive to a variety of noises, such as loud bangs, clapping or laughter, and they may become tense and fearful when hearing or anticipating these sounds.

Of course every child is different, and not all children with WS will show all of these characteristics to the same extent. However, research undertaken over the last few years with children and adults with WS indicates that these difficulties are common to many of them, and that unless parents tackle them early on they may well persist into adulthood. We would therefore encourage parents and teachers to address these difficulties as early as possible in the child's life. In the following pages we describe these difficulties in greater detail and suggest ways in which you can guide your child in each area. The methods and approaches we will describe are well tried and have been used successfully to cope with the difficulties of a wide variety of children. They can be adapted for use with any individual child. Some examples focus on boys, some on girls, but only because we wanted to avoid using "he/she" all the time. The tips work equally well with children of either sex.

FEEDING AND EATING DIFFICULTIES

Feeding difficulties, including vomiting and refusal to feed, usually appear in the first months or year of life and are one of the earliest symptoms of the condition. If a blood test shows the child to have a high level of calcium, he may be put on a low-calcium and Vitamin-D restricted diet, and a low-calcium milk preparation (Locasol) may be recommended. With time, the feeding difficulties will lessen and disappear.

In some cases, problems with swallowing and chewing persist and you may find it difficult to feed your child with anything other than a soft food diet. And a child who only takes smooth foods will never learn to chew! In this case you could try to introduce first very small and then larger lumps into the smooth, liquidised food, or gradually to make the food thicker. Then introduce a few small pieces of minced meat etc. into the soft food, and move on to easily chewed foods like fish fingers and rissoles. You may want to enlist professional help from a psychologist or speech therapist (through your Paediatrician or local Child Development or Child Health Centre) to introduce a feeding training programme, with emphasis on developing sucking, swallowing and chewing skills.

Once the child's blood calcium has stabilised over a period of time, the Paediatrician will advise you about taking your child off the low-calcium diet. For many of the children feeding will now no longer be a problem. However, some children continue to be fussy eaters and may refuse to eat all but a very limited range of foods even once they are off their diets. Such idiosyncratic diets can cause difficulties, for example when the child starts school and is expected to eat set school meals. Since at this stage there is no medical reason for a limited diet, your child should be encouraged to try and eat a wider range of foods. One approach you could use is to insist, at mealtimes, that the child try just one spoonful of a food he normally will not eat, and then immediately praise him and reward him with his favourite food, and perhaps also with a favourite toy. Such an approach fits in with our knowledge of the principles of rewarding desirable behaviours. By immediately following a desirable behaviour with a reward, you make it more likely that this behaviour will occur again. Gradually, you should require the child to eat larger amounts of the non-preferred foods before giving him preferred foods and other rewards. Sooner or later most children actually begin to enjoy eating these other foods, and forget their earlier aversion to them.

A few children develop a habit of vomiting at mealtimes if they are made to eat foods they do not like, or perhaps as a means of gaining attention from parents, or when they are anxious or under pressure. If it is confirmed that there is no physical reason for the vomiting, it is advisable for parents to be firm and to withdraw sympathy and attention at times when the child vomits (and perhaps to insist on the child cleaning the mess himself if he is able to), but to praise the child and reward him with toys or with foods he likes if he does not vomit at mealtimes or at other times during the day. Another approach is to take the pressure off mealtimes and leave the child to eat as much or as little as he wants, on the theory that he will eat if he is hungry. This approach will obviously only be effective if the child is denied sweets, biscuits and snacks between meals. Some children dribble, and they will need regular reminders from adults to close their mouth and swallow. Again, a reward system may serve as an added incentive, with the child being rewarded if he can spend increasing periods of time without dribbling.

SLEEPING DIFFICULTIES

Some children with WS have problems settling down to sleep, or they may wake in the night and cry or go to their parents' bed. These problems are by no means unique to WS, and as with other children, they can become severe and prolonged and parents could find themselves spending several hours each night comforting their child or taking her into their own bed. Dealing with these difficulties requires a firm and consistent approach. If your child takes a long time to fall asleep and continually calls out to you or insists on your presence before falling asleep, try and set up a fixed bedtime routine. Once you have said firmly that it is time to go to bed (preferably at a fixed time each night), help her to get ready for bed and settle down. You may want to read a story or sing a song, and then say firmly that it is time to go to sleep and that you will be leaving the room. You may want to give a favourite toy for reassurance, or leave a light on in the room, and perhaps use a dimmer switch, which can be faded gradually as she gets used to it. Once you have left the child, you should try and be consistent about not answering calls or cries and not going back into the bedroom. To begin with, the period spent ignoring your child's crying and calling out may seem like an eternity, but if you time it you will probably find that it is a lot shorter than you think, and it will become progressively less over time. If the child gets out of bed and comes to you, firmly return her to bed each time and leave the room as soon as possible.

Praising your child for desired behaviour and giving detailed feedback about her behaviour is important. If the child is old enough to understand, you could introduce a star chart and stick a paper star or sticker onto the chart each morning as a reward for settling without problems the previous night. In this way, both you and your child will have a record of the progress that is being made over time.

If your child wakes during the night and cries or calls out to you, again insist that she goes back to sleep, once you have made sure that she is all right. Ignore any further calls. If she comes into your room, firmly take her back to her bed and leave the room as quickly as possible.

When coping with sleeping difficulties, it is important to be firm and consistent and not to give in to your child's demands. This is not always easy to do! Some of the approaches described above are easier to use with children who already have language. With younger children and infants, other methods may be needed. If the sleeping problems persist, you may want to approach your health visitor, GP or Paediatrician for advice, or ask for a referral to a Sleep Clinic or the local Child Clinical Psychology Service or Child & Family Mental Health Service (CAMHS). The Royal College of Psychiatrists also publishes a leaflet on sleep problems with suggestions for further reading and audiotapes, etc (www.rcpsych.ac.uk/publications).

Some parents have also found a milk drink with high melatonin levels ("Night Time Milk" obtainable in big supermarkets) very helpful.

TOILET TRAINING

Wetting and soiling can often persist in the pre-school years and into the school years; or they may appear as new problems, and may be exacerbated by anxiety or excitement. With a well worked out programme most children can become toilet trained well before they go to school, or helped to become clean or dry again if they have regressed.

Most children become toilet trained through a combination of imitation, parental pleasure and displeasure, and luck. But if a child is not making progress in toilet training in the usual way, then perhaps by age 3, 4 or 5, parents might decide to try a more systematic approach to toilet training. When you decide to toilet train your child, take him out of nappies. To start with, take him to the toilet at regular intervals (every half hour or hour) and encourage him to sit on the toilet or pot for several minutes. Reward him for performing on the toilet with praise and perhaps with a favourite toy, or a star on a star chart. Using a musical potty may increase the child's interest in performing on the pot and may also serve to focus his attention on what he is doing. If the child has an accident, show him that this is not desirable, but without being punitive. You may also want to make him help you to clean and wash his pants. If this approach is successful and the child does not wet or soil his pants in between, gradually increase the time interval between toileting. Teach him to say the word "toilet" or a related word or gesture that indicates his need to go to the toilet. With time most children will come to indicate or take themselves off to the toilet when they have to go.

The above habit training method may not work for every child, and if you find no reduction in the number of accidents after giving the approach a fair trial (which may be several weeks or months), other methods may be needed. One such method is a pants alarm. This consists of a sensor, which is attached to a pad that is inserted into the child's pants. The sensor is attached to a small alarm, which can be pinned on to the child's clothing. If the child begins to wet the pad, the alarm goes off. The child is then taken, or reminded to go, to the toilet to finish urinating. This technique helps the child to gain greater awareness, and hence greater control of his bladder function, and it has been successfully used with many children of different ages. Parents may be concerned that children with WS who are sensitive to noise will become upset by the alarm. In fact we know of a number of WS children who have used this device without difficulty. But if you fear that the noise of the alarm may be a problem for your child, allow him to play with the alarm and hear it over a number of days so that he can get used to it before you use it for toilet training. Your health visitor or GP can refer you to a local clinic or to a Psychologist who can provide you with a pants alarm and guidance on how to use it, or suggest other approaches to try. Bear in mind that urinary tract infections can cause, as well as be caused by, episodes of wetting, and should always be checked for by the GP. In addition, it is important to note that children with WS often need to urinate more frequently than other children, and they should be given the opportunity to go to the toilet as often as needed.

If your child wets the bed at night, you could in the first instance introduce a star chart, whereby you reward him with a star and lots of praise if he has not wet the bed the previous night. This approach can be very effective, particularly if combined with a programme of lifting the child to the toilet several hours after he has fallen asleep. If he can remain dry, you can gradually bring the time of lifting him forward until it coincides with his bedtime. No approach will work immediately, and it may well take several weeks or months for such a strategy to start taking effect. Another approach to bedwetting is to use a bedwetting alarm (see above). If the child begins to wet the pad the alarm will go off, which causes the child to wake up and stop urinating.

The parent should then take him to the toilet and reset the alarm. Rewards may be used in conjunction with the alarm, and the child rewarded with praise and toys or stars for dry nights.

Teaching bladder control will sometimes indirectly result in bowel control being achieved. If soiling continues to be a problem, then over several weeks or months, encourage the child to sit on the toilet or pot for several minutes or longer at the time(s) when he is most likely to pass a motion (e.g. after mealtimes), and use a combination of praise and other rewards when he does open his bowels in the toilet or pot. Try not to get angry if he soils in his pants, but indicate that this is not desirable. You may also require him to help you to clean himself and wash his soiled clothing. Related behaviours that may need to be taught include dressing and undressing, the ability to wipe clean afterwards (moist toilet tissues can be a help here), flushing the toilet and hand washing. Constipation may be a complicating factor in soiling, in which case it will be particularly important to ensure that the child eats a balanced diet with plenty of fibre, fruit, vegetables and liquids. The GP or Paediatrician may need to examine your child to check whether he is constipated; in this event the rectum and colon may have become impacted with hardened faeces, and it would be necessary for the impacted mass of faeces to be removed before bowel training can proceed. Subsequently medication can help to soften stools so that the problem is less likely to recur.

You may find it helpful to seek advice and support from a health visitor, Child Psychologist or other child health professional, on the most appropriate method to use with your child in toilet training. Furthermore, if he is attending a nursery, playgroup or school at this time, invite the staff there to cooperate with you on toilet training so that a consistent training approach can be adopted.

The Royal College of Psychiatrists also produces a helpful leaflet and recommended reading list to help parents (www.rpsych.ac.uk/publications).

DRESSING

The tasks of dressing and undressing require muscle co-ordination and planning, and this is something children with WS may find difficult to master. The child may need help with putting on clothes, doing up buttons and shoelaces etc. Because there is usually so little time in the morning, parents often find it more convenient to dress the child completely, in order to save valuable time. This can become habitual, not just with a child with WS but with any young child. However, the child needs to be encouraged to independence, to learn to do things for herself. Thus, over time you can slowly encourage her to do more of the dressing independently; for example, teach her to put on her own pants and then help with the buttons or zip.

Learning to dress can be practiced at other times of the day, and not just during the fraught morning period when time is of the essence. Clothes should be as easy as possible to put on. Elasticated waistbands, loose fitting sleeves and necklines, Velcro or popper fastenings, slip on shoes, all help to reduce hassle and increase the child's independence. Colour coding, too, is helpful. For example, putting a coloured mark or sticker inside every *right* shoe (and even putting a dab of same colour nail varnish on the big toe of the same foot) can help shoes to go on the correct foot. A coloured tab sewn into the *front* of jumpers or vests etc, can prevent clothes going on back to front.

It is helpful to break down tasks like dressing into smaller steps and teach the child one step at a time. For example, if you are working on teaching her to put on her socks, you might first put on each sock up to the ankle and teach the child to pull up the sock from the ankle. Once she is able

to perform this step correctly, go on to the next step -put the sock on her foot half over the heel and teach her to pull the sock over the rest of the heel and up. The next step is to put the sock on the foot to the heel, then on to the instep, then just over the toes -and get the child to pull the sock up over more of the foot at each step. Eventually, you will be able to hand the socks to the child, and then leave them on the bed or chair, so that she can take each sock in turn, put it on her foot the right way round and pull it up. Learning to do up buttons can sometimes be helped by encouraging the child to practice on buttons 'off the body' first. Cut some buttons and button-holes off an old shirt or cardigan and get your child to practice on these. Giving the child verbal cues is also helpful. As an example, when teaching her to do up shoelaces, she can be helped to talk herself through the routine, for example by saying out loud 'cross the laces over' 'pull one through' 'pull tight' as the child performs each step. In this way she learns the words and can then prompt herself in the task.

SOCIAL RELATIONSHIPS

Some children with WS find it difficult to make friends of their own age and show no interest in playing and interacting with their peers. As they become young adults difficulties in establishing and maintaining friendships with others of their own age may become even more apparent, and may be a source of frustration to them and to their families. On the other hand, they love the company of adults, are affectionate and eager to please, and will often seek out adults to engage in conversation. At the same time, they often lack understanding of the underlying, 'unwritten' rules governing social interactions and fail to recognise the social constraints that are apparent to others. Thus, they may approach strangers in an over-friendly and over-familiar manner and will often tag along with them. Understandably, this can be a major worry for parents, who fear that the child is too trusting and could be taken advantage of if he is not watched and supervised all the time.

Adult friends and relations do not always interact appropriately with children with WS, and they may on occasion 'over-indulge' or 'baby' them. It is important to try and educate adult friends and relatives to behave appropriately with your child, but also to show some tolerance and patience for any unpredictable behaviour.

Helping children with WS to make and maintain friendships with people of their own age is a complex issue. Particularly as the children move into adolescence, they may be reluctant to socialise with other people with special needs, but they may also be unable to make friends with 'normal' adolescents who may have little patience with them. If your child is young, you could help by inviting children from school to your house and initiating and supervising games for them to play. These could include board games, ball games and make-believe play with dolls, cars, etc. Parents and school staff can work on teaching the children social skills, including turn taking and sharing. Many parents also find that their children integrate very successfully into organised and supervised children's activity groups such as Scouts or Guides, which often appeal greatly to their gregarious and extrovert natures.

You may need to be actively involved in organising your child's leisure time even when he is older, since the initiative will not necessarily come from him. Many parents find that if they do not actively organise outings and activities for their older children, they will be satisfied to stay at home and watch television or listen to music. Information about local social and sports clubs for children and adults with a range of disabilities and learning difficulties may be available from your child's school, or from the local Child Development Centre or Social Services Department, or from a health visitor. These clubs often arrange supervised outings, sporting and other social activities where the person with WS can meet others on a regular basis.

A number of organisations arrange supervised holidays for children and adults with disabilities, and financial assistance for such holidays is often available. You can find out about these from your local Social Services Department or from your child's school. The Williams Syndrome Foundation, too, provides annual holidays for children and adults with WS (both with and without their families).

Some families make use of short-term residential or respite care for their child arranged by local Social Services Departments. Under this scheme the child can spend a few days or weeks with another family, or in a local residential hostel or unit, at regular intervals, thereby giving parents a break, as well as affording the child the opportunity to meet new people and enjoy new experiences. Parents can obtain information on what is available locally from their Social Services Department, but need to bear in mind that the type and availability of respite care services is very variable from region to region.

It is important to stress that while some people with WS have difficulties in establishing relationships with peers, others do not. On the contrary, with their friendly and extrovert natures and also their sensitivity and concern for others, many children and adults with WS are popular and well-liked, and establish warm friendships with people outside their immediate families.

SUGGESTIONS FOR DEALING WITH THE WS CHILD'S OVER-FRIENDLINESS

It may seem unfair to put limits on the child's friendly overtures to adults, but if this way of interacting with strangers is encouraged, it could lead to serious problems at a later date. By looking ahead and anticipating future needs, many crises can be avoided.

- 1. When the child meets someone he knows, or when he is introduced to a new person, he should be TAUGHT how to GREET the person in an appropriate way and discouraged from hugging or kissing the person.**
- 2. Social situations can be REHEARSED with the child at school and at home. For example, you could pretend to be an adult friend and allow the child to practice greeting you. MODEL appropriate greetings for the child and instruct him to IMITATE you. PRAISE him for appropriate behaviour.**
- 3. Children can be taught simple rules, such as not to stand too close to others and not to stare; VIDEO and ROLE PLAY techniques can be useful here.**
- 4. Parents and teachers should start as early as possible to DISCOURAGE the child from approaching people he does not know. Whenever he does approach a stranger, parents and carers should indicate their displeasure and make it very clear that this is not allowed.**

EMOTIONAL/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.

ANXIETY

Children with WS are often described as being over-anxious and easily upset by criticism and frustration. They may worry excessively about the health and well being of their families, themselves and even strangers, as well as about unfamiliar situations and all kinds of imagined disasters. This concern and sensitivity for the needs of others can be an endearing aspect of their personality, which makes people warm to them. But it may also mean that the child with WS frequently demands attention and seeks reassurance from the people around her. It is advisable for parents and other adults to strike a balance between comforting and reassuring the child in such circumstances but not making too much fuss, since this may well encourage and exacerbate feelings of upset and worry. You may find that you are spending a great deal of time comforting your child, but with no decrease in her level of expressed anxiety. If this is the case, you might decide to adopt the following strategy:

- **Put a fixed time limit on the time you spend comforting the child. For example, you might agree to spend one or two minutes reassuring her if she expresses anxiety.**
- **Then move on to another topic of conversation or activity.**
- **In this way the adult acknowledges the child's anxiety and provides comfort and reassurance, but also circumscribes it within time limits so that it does not become just another way of gaining unlimited adult attention.**

Because of their sensitivity, people with WS may be very susceptible to stress and to the demands of others, and may find it very difficult to cope in environments that are excessively demanding. If you feel that your child is becoming more nervous or anxious than usual, then it will be important to examine the home and school environments to ensure that the demands being placed on the child are not excessive. In general, children with WS do best with a predictable schedule and a set routine, and benefit from preparation before changes in activities or in routine. Stress and anticipatory anxiety can often be reduced by spending a brief period ahead of time preparing the child for a change or for a difficult task or event, enumerating the difficulties to be faced and talking through possible outcomes.

PREOCCUPATIONS, OBSESSIONS & STEREOTYPED BEHAVIOURS

Children and adolescents with WS often display intense fascinations and preoccupations with certain objects (such as insects, cars, or electrical gadgets), particular topics (disasters and violence on the news, illness, future events such as birthdays and holidays etc.) or certain people (a particular member of the school staff, a television star or neighbour). They may spend a great deal of time absorbed with these topics and talk about them repetitively. It is best if you can nip such obsessions in the bud before they get a real hold; so if you suspect that a particular

behaviour is becoming obsessive, try to divert attention elsewhere, and introduce new activities and interests for the child. If the preoccupations are already well established try to circumscribe them to keep them within acceptable bounds. For example, where a child is very preoccupied with a certain toy, or becomes attached exclusively to one other child and always wants to stay close to him, the school staff might decide to allow him to play with the child/object in question for a fixed time (say 10 minutes every day or two) but only if he controls his preoccupation in between these times. If unable to do this he would forfeit some of his 'special time' with the child/object. The aim is to bring the preoccupation under control and gradually to reduce the amount of time spent in this way. Alternatively, time spent with the object or person the child is preoccupied by could be used as a reward for desired behaviour in other areas. For example, the child might be awarded stars on a star chart for completing arithmetic assignments, for concentrating in class, for finishing his dinner etc. And once he has earned a pre-agreed number of stars he can be rewarded with an opportunity to play with his favourite friend or toy. The obsessional interest or behaviour could also be channelled into useful activity, for example practising pencil control by drawing pictures or writing stories about a favourite topic.

Some individuals with WS display stereotyped behaviours such as rocking or hand flapping. These behaviours, which may also be displayed by other people with learning difficulties, may be particularly prominent when the individual is anxious or agitated, when he is bored, or alternatively when he is absorbed in a particular task or trying to concentrate on something. In some cases rocking may actually aid concentration. However, in general it is advisable to make the child aware of his rocking or hand lapping as soon as it begins, and to ask him to stop. He will usually need frequent reminders to start with, although with time a brief nudge or pre-agreed word will usually suffice. It is often helpful to combine this approach with a reward system whereby the child is systematically rewarded if he can spend increasing periods of time without rocking. Encouraging the child to change his sitting position or the type of chair he usually sits in (when watching television, for example) may also be effective.

TEMPER TANTRUMS AND AGGRESSIVE BEHAVIOURS

Children with WS are mostly even-tempered and co-operative. But like other children they may have severe temper tantrums when they are frustrated, when they cannot make their needs or wants understood, when they want attention or cannot get their own way. Tantrums and aggressive behaviour towards others can occur in both young and older children. And, as any parent knows, they are by no means confined to children with learning difficulties.

You should observe the child carefully when she has a tantrum and try to identify the situations and events that trigger the tantrums. For example, if you find that fear of particular events or loud noises or provocation by another person triggers a tantrum, then you could try and anticipate these and divert the child's attention onto something else before the tantrum develops. You could also teach the child more appropriate ways of communicating with others to indicate her needs and wants, which would make it unnecessary for her to throw a tantrum. When faced with tantrums or other unacceptable behaviours you and other family members might agree to ignore the child and turn away from her for a short period. As soon as she stops misbehaving you can attend to her again. Ignoring can be an effective way of managing most difficult behaviours, except where the child is being destructive or is likely to hurt herself or someone else. In such cases an effective approach is to remove the child, for example by putting her in the hall or bedroom, and to make it clear that she cannot rejoin you and the rest of the family until she has calmed down. It is important for the adult to try and remain calm and to adopt exactly the same approach every time the child has a tantrum, since an inconsistent approach will only confuse her. As soon as she is quiet, go and praise her for having quietened

down and allow her to rejoin the family. At first there might be a temporary increase in the disruptive behaviour. But if you insist that the child remains in her room and ignore her until she is quiet, she will soon come to understand that the tantrums do not enable her to get her own way, and their frequency and severity will decrease.

The same approach can be used to bring other undesirable behaviours, such as hitting other children, biting, and breaking things, under control. Moodiness can be a feature of the emotional liability of some children with WS, and it can become particularly prominent in adolescence. It may be characterised by sullenness, muttering under one's breath or by overt defiance. Here too, if distracting the individual is not immediately effective, it may be advisable to make it clear to her that moody behaviour is unacceptable and that she will be expected to go out into the corridor or to another room until she has regained her good spirits and is ready to cooperate. In this way the adult withdraws attention from the child when she is moody and uncooperative, but rewards her with praise and attention when she is more cooperative. Again, it is important to stress that moodiness is not an invariable feature of the syndrome, and many WS individuals are by nature just the opposite -friendly, caring, helpful, and eager to please. However, if you feel that aspects of your child's behaviour are getting beyond your control, you can obtain professional help from a local Child Clinical Psychology Service or Child and Family Mental Health Services (CAMHS), via your health visitor, GP, Child Development Centre or your child's school.

HYPERSENSITIVITY TO SOUNDS

About 90% of children with WS are hypersensitive to particular sounds that would not cause discomfort in most people. These can include sudden loud noises like thunder, objects falling or balloons bursting, hand clapping, electrical noises like vacuum cleaners, drills and electric shavers, loud music, and in some cases even people's voices or laughter. This phenomenon is termed 'hyperacusis'. The noises may be very distressing to the children, who will typically put their hands over their ears and cry, or try to avoid the sounds, for example by leaving the room or turning off the television or radio. Noisy and rowdy environments may be particularly upsetting. In some cases the hyperacusis becomes less of a problem in the child's teens or in adulthood, but in other cases it continues to cause difficulties.

The reason for this sensitivity to noise is not yet known, but it is likely to be related to the fact that individuals with WS have lower thresholds for sounds than do other people. It is possible that the noises may be physically painful to the ears, or they may simply startle the child.

Some people recommend the use of earplugs to block out noise. Since earplugs are likely to block out important sounds (like speech) as well as distressing sounds, great care should be taken in their use. If you do decide to let your child use earplugs, they should only be used for a brief time where a particular noise is causing the child distress. And don't forget to take the earplugs out immediately thereafter!

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 helps.**
- 2. Whenever possible provide a warning just before predictable noises (e.g. before fire drills or before switching on the food processor).**
- 3. The reactions will often diminish if the child is able to exercise some control over the sounds that cause discomfort. For example, encourage him to use the vacuum cleaner, or to help with household chores by turning on the washing machine or food mixer.**
- 4. The child may be reassured if he knows that he can leave the room for a few minutes at any point, if he is exposed to a distressing noise.**
- 5. Repeated gentle exposure to the sound may help the child to get used to it. For example, tape record one or more distressing sounds and encourage him to play back the tape, quietly at first, then gradually increasing the volume.**

SPEECH AND LANGUAGE SKILLS

In early childhood, children with WS may be very slow to develop speech. At this stage, verbal stimulation is vital. Incorporate language into play, talk to your child while you are dressing, washing and feeding her, and encourage her to respond to you with babble and vocalisations. In addition, you might find it valuable to spend 5 to 10 minutes each day working with your child on building up her vocabulary.

Depending on her language level, you could choose 3 or 4 of the most useful nouns, verbs or prepositions to work on at a time. For example, if you decide to work on object names, choose 2 objects (e.g. cup, ball) and ask 'What's this?', pointing to one of the items. If the child does not respond, prompt her with 'It's a ball/cup' and encourage her to imitate you. When the child says the word, praise her and give her the object to play with for a short time. At first, any attempt that sounds like the word should be rewarded, but over time, require the child to approximate the word more closely. Gradually introduce more objects and also pictures of objects and then verbs, adjectives, prepositions, etc.

To teach word comprehension, you could start by placing 2 or 3 objects in front of the child and say 'Give me the ball/cup'. Praise the child if she is correct and allow her to play with the object

for a short while. If the child does not respond correctly, prompt her to pick out the right object. Again, you can progress to teaching her to identify other objects, and then to respond to a range of commands (e.g. sit, stand, jump, wave) with the appropriate actions. Once she can reliably follow simple commands, you can introduce more complex instructions that are of help in daily living. These might include instructions such as 'Fetch your coat' and 'Go to the toilet'. Each stage of the training procedure should follow the same pattern, with physical guidance and prompting, and also praise and other rewards being used to encourage motivation. When showing your child how to play with toys that interest her, you should use simple language to accompany the play, and then encourage her to take her turn. As she takes a turn imitating your play, it is likely that she will also try to imitate the language spoken.

Children with WS benefit from speech and language therapy at all stages of their development, and the therapist should also work closely with parents and provide suggestions on activities you can carry out at home in order to expand the child's language comprehension, spoken language, oral motor abilities and speech production. In the case of children whose speech is particularly slow to develop in the pre-school years, speech and language therapists may recommend introducing an augmentative communication system such as manual signing (e.g. Makaton) to support and encourage speech development.

DISCOURAGING EXCESSIVE CHATTER AND INAPPROPRIATE SPEECH

If a child has a favourite topic she talks about repetitively (e.g. trains, illness), or set questions she always asks, it may be advisable for you and other adults to IGNORE her when she launches into her 'favourite' topic. If you answer such 'pet' questions each time, the child will be encouraged to ask them again and again!

Your family and friends could make a rule to answer the child's questions ONCE but to IGNORE all repetitive questions and topics thereafter, and either to turn away from the child or to CHANGE THE SUBJECT straight away and move on to topics that are more relevant.

Another useful strategy is to insist, when a child's question has been answered and she asks it once again, that she repeat the answer that has already been given.

A CONSISTENT approach is important, so urge all adults to adopt exactly the same strategy at home and also at school. With such methods, socially appropriate speech should increase, and inappropriate speech will steadily decline.

By school age, most children with WS will have developed fluent, articulate speech with sophisticated, wide ranging vocabularies and complex and grammatically correct sentences, which are quite out of keeping with their general level of learning difficulty. They may use long words and unusual phrases which they have heard other people use and remembered, and they may chatter to anyone they meet, if given the chance. But often their speech is inappropriate and repetitive. They will perseverate on certain favourite topics of conversation and talk about these again and again, much to the irritation of family and friends.

Many use chatter and incessant questions to initiate and maintain social contact with other people, and they are adept at using their superior verbal skills to get attention. Often favourite topics are simply areas the child is confident discussing, and is relying on in order to take part in the conversation. It is important to encourage the child to talk to people she meets, but on the other hand you will want to discourage her from chattering too much in company and from asking incessant and irrelevant questions.

Some children and older individuals with WS echo or repeat phrases and sentences spoken by the person they are interacting with, often with little understanding of what the person is saying. If the echoing is caused by poor comprehension, be sure to simplify your speech. However, if the child's echoing is simply a habit he has developed, then ignoring it and distracting the child by moving on to another topic can be a useful method of discouraging such meaningless utterances.

Understanding of language is in many cases more limited than the child's expressive language might indicate. If your child responds to your comments and questions inappropriately or irrelevantly, this may be because she does not fully understand you. You may need to simplify your conversation and to make sure that your child understands what you and other people are saying.

If you are worried about her language development or communication you can ask for her progress to be reviewed by a speech and language therapist, either through the school or through your child's Paediatrician, GP or health visitor.

CONCENTRATION DIFFICULTIES AND OVERACTIVITY

Poor concentration, distractibility, restlessness and overactivity are highly characteristic of children with WS. Where the children are also hypersensitive to noise, they may be particularly distracted by incidental and irrelevant sounds at times when they are required to concentrate, for example in the classroom, or when they are being given instructions or listening to a story. Clear and deliberate instructions to the child will help him to understand what is expected, and regular prompting and reminders will help him stay on the task and remember what he has to do.

Where the child is fascinated or obsessed by particular topics or objects, then books or games geared to these can also be used to encourage concentration, at least initially. Alternatively, you could use something the child is obsessed with (e.g. a car or machine) as a reward he can play with, but only after he has first spent a set period of time engaged in more structured work. So you might, for example, make an agreement with your child that after he has spent 5 minutes in a structured activity he can engage in his favourite preoccupation/obsession for 2 minutes. This may be playing with a favourite toy, watching the builder working next door, or chatting to you.

This 2 minute break would then be followed by a further 5 minute period of structured work, and so on. It is also helpful to break down tasks into short, simple steps with cues -such as pictures or words -to indicate clearly what needs to be completed at each stage.

In adolescents and adults with WS, overactivity is no longer a prominent feature, but a limited attention span may continue to be a problem.

Some children benefit from learning to talk out loud to themselves, repeating an instruction or talking their way through an activity. This may be particularly useful as an aid to concentration for older children and adults. Thus you could teach the child to remind himself to keep working on the task at hand, initially by thinking aloud and later by silent instructions to himself. So, while working, he is taught to tell himself at regular intervals to 'keep on working', to 'concentrate and look at what I am doing' etc.

Many of the children are also impulsive and find it difficult to take turns and wait for an adult's attention. They may repeatedly interrupt adults' conversations because of their desire for attention. Again, you can reward the child if he is able to sit quietly and wait his turn, initially for a few moments, and then for longer periods of time. One parent found that on long journeys, when continual chatter and interruptions can be particularly tiring, a 'sponsored silence' was extremely effective. A piece of fruit, a sweet or small coin offered for a carefully timed 10 minute spell of silence can work wonders for the frayed nerves of the whole family!

It is useful to remember, and to point out to teachers, that because children with WS may be easily distracted by what is going on around them (even more so than many other children with learning difficulties), they are likely to pay attention and work better in settings that are as quiet and free from distractions as possible. Short periods of work interspersed with frequent breaks will also be necessary. Most children need one-to-one guidance and attention from an adult in order to complete a task, at least initially. Working with the child on his own or with a few other children, in a quiet room, and for short periods of time, is likely to be the most appropriate.

IMPROVING THE CHILD'S CONCENTRATION SPAN

- 1. Reward the child with praise or toys for sitting still and listening, or for engaging in some constructive activity such as looking at a book or doing a puzzle, for progressively longer periods of time.**
- 2. Initially require him to sit still and pay attention for 1 or 2 minutes, and immediately thereafter reward him with a favorite toy or activity and praise. Adult attention and conversation are likely to be particularly effective reinforcers.**
- 3. Gradually, through a series of stages, build up to work periods of 5, 10 and then 15 minutes.**

SCHOOL RELATED CONCERNS

a) Type of Schooling

Children with WS have an unusual pattern of abilities, often with relatively good spoken language but poorer perceptual and motor skills and concentration difficulties. As a result, they have special educational needs that are different from those of other children, and it can be quite difficult to find a school that will be exactly suited to their particular learning needs. Their verbal skills may suggest a higher level of ability, but if they are placed in a mainstream school or in a school for children with Moderate Learning Difficulties, they may be unable to cope because of their lower abilities in non-verbal areas and their limited concentration. There is no one type of school that is ideally suited to the needs of the child with WS. Education Authorities differ greatly in the way they allocate resources to children with special needs; finding the most appropriate school will depend on the individual child's level of ability and profile of strengths and weaknesses, and also on the provisions in the particular schools that are available locally.

Some children start off in a mainstream school and cope very well there at the primary level, though typically with additional educational support being provided for them. They may get help from a support teacher or assistant for some part of each school day or for a few hours per week, for example in maths, reading and writing. But for other lessons and at playtime, the children are with their ordinary classes. At the secondary level, some of these children move to special schools or units where they can benefit from more individualised teaching and learn at a slower and less pressurised pace. A few children continue in mainstream at the secondary level, for example attending a remedial class for some lessons and the ordinary class for other lessons. This gives them the opportunity of integrating with their peers on the playground and in certain lessons.

Mainstream schooling may not be appropriate for all or even the majority of children with WS. Many of the children need the slower pace and more individualised attention that are currently provided in special schools and units. Each child's needs will be different. In a study we undertook in the late 1980's we found that:

- **30% attended schools for children with Severe Learning Difficulties**
- **30% attended schools for children with Moderate Learning Difficulties**
- **30% attended schools for children with complex special education needs**
- **10% were in mainstream schools (with support)**

With the greater emphasis that is now being placed on integration and inclusion, we are seeing increasing numbers of children with learning difficulties, and thus also more children with WS, being educated in mainstream schools, with varying amounts of additional support. We believe that for inclusion to work effectively for children with WS, classroom settings, which offer a great deal of flexibility and substantial support, will be necessary. Typically this will require:

- **extra adults in the classroom**
- **small classes**
- **intensive involvement of specialist teachers**
- **learning support assistants**
- **carefully structured and graded work plans and Individual Education Plans (IEPs)**
- **teamwork between classroom teacher, assistant, specialist teachers and speech and occupational therapists**

Under the 1996 Education Act and 2001 SEN and Disability Act Local Education Authorities have duties to identify, assess and provide appropriate education for all children with special needs from the age of 2 to the end of compulsory schooling at 16 -and up to 19 if the student wants to remain in school. You, as parents, have the right to ask the Local Authority to initiate a Formal Assessment of your child's Special Educational Needs from the age of 2 or at any point in his school career, in order to ensure that his special educational needs will be identified and met. Alternatively, your child's school may feel that he has special educational needs and they may ask the Local Authority for an Assessment of these needs to be undertaken. Where professionals working in the Health Service feel that a child may have special educational needs they must inform the parents and, after discussion with the parents, the Local Authority.

Once the Assessment is underway, your child will be assessed by an educational psychologist to determine his level of ability and specific needs, and reports will also be obtained from the school (head and teachers), school doctor or paediatrician, and any other professional who is working with your child (e.g. speech therapists, physiotherapist, occupational therapist, clinical psychologist). You will be sent copies of these reports. You too will be invited to submit a report (Parental Advice Appendix A) to the assessment panel, stating your views of your child's abilities and needs, and the type of schooling you feel would be appropriate. In such a report, you will want to point out your child's particular difficulties and special needs, but also his special strengths and the skills and tasks he excels in. For example, many WS children have very good verbal and social skills and extremely good memorising abilities. They may love talking to and interacting with others and respond very well in settings which provide them with a great deal of verbal and social stimulation. Since most professionals will know very little at all about the special abilities, difficulties and needs of children with WS, you may find it useful to submit, along with your reports, additional information about Williams Syndrome, including a copy of this booklet. You will have a chance to visit schools that may be considered for your child, and you have the right to express a preference for a particular school which you feel would be most suitable. However, it is the duty of the Local Authority to agree to the school named in the statement. You will want to try and make sure that the type of school being considered for your child can cater for his special needs but can also nurture and stimulate his special skills and talents.

Once the information has been gathered from the professionals working with your child and from yourselves, the Local Authority will recommend how and where the child's special educational needs should be met. The abilities and needs of children with WS are so complex that in our opinion most will need the protection of a Statement of their Special Educational Needs. This is a legally binding document drawn up by the Local Authority, which describes the child's special needs and specifies what provision the child should have to support him in his learning. However, the Local Authority may decide that the child's needs can be met without a Statement being issued. If you disagree with the Local Authority's decision not to issue a Statement, or with the proposed content of the Statement, and cannot come to an agreement even after further discussion with the Authority, you can appeal against the recommendations to the independent Special Educational Needs and Disability Tribunal.

Once a Statement has been made, it must be reviewed annually. Furthermore, the first Annual Review after your child's fourteenth birthday must introduce a Transition Plan, which will help in planning for his transition to adult life and continuing education.

Advice and support on the Statementing process, including how to prepare your report and how to go about lodging an appeal, can be obtained from your child's school, educational psychologist and from officers within your Local Authority. Parents can benefit from the support of an independent Named Person -someone who is knowledgeable in the statementing process and can help you through it; there are also voluntary organisations and Parent Partnership schemes in many areas who offer independent advice and support to parents. The Williams Syndrome

Foundation has produced a leaflet 'Profile of Special Educational Needs for Children with Williams Syndrome', and can be contacted directly for advice in individual cases.

b) Working with Teachers

Most teachers will never have taught a child with WS before, so they will know very little about the syndrome. Teachers frequently say that they find children with WS confusing and puzzling to teach because their relatively good verbal abilities, engaging personalities and sociability can result in an overestimation of their overall abilities. We have found that teachers are very keen to find out about the special needs and difficulties of children with the condition, and both they and your child will benefit the more they learn about WS. The Williams Syndrome Foundation has a number of articles and leaflets that would be helpful for school and nursery teachers, including:

Udwin, O. Howlin P and Yule, W. (2007)
Williams Syndrome: Guidelines for Teachers.
Published by the Williams Syndrome Foundation.

We strongly recommend that you pass on copies of this booklet and other articles to your child's teachers and encourage them to read them.

c) Perceptual and Motor Difficulties

Many children with WS have particular difficulties with gross and fine motor co-ordination, with discrimination tasks, with orienting themselves and objects in space, and judging distances and directions. These children will need help and opportunities for repeated practice of tasks in these areas, both at school and at home. Activities such as swimming, horse riding and gym help to improve co-ordination and visuospatial skills and are popular with many children with WS. Tasks such as sorting and matching objects and shapes, drawing and copying shapes and being able to trace over lines, are all important skills to master before the child can move on to the basics of reading and writing. Such tasks may be more difficult for children with WS because of their difficulties with visual perception and motor co-ordination. They will need a lot of practice in all these areas, and you may want to practice some of these exercises with your child at home before he starts school.

Because many of the children tend to have good spoken language, we find that it often helps if they can be encouraged to talk themselves through each step of an exercise while they are doing it. This can help them to focus their attention on the tasks and also provide verbal reinforcement and support for the activities.

Some parents and teachers have also found it helpful to introduce teaching materials using objects and themes that the child is especially interested in or even obsessed with, in order to motivate her to work on tasks which she may not be directly interested in. For example, if the child is fascinated by cars or machines, get her to trace over, copy and draw outlines of these. If the child loves music and singing, try and incorporate these into the tasks. Parents can do a variety of activities with their child to help her develop the fine motor skills necessary for writing, including dot-to-dot drawing, mazes, colouring books, clay, and other activities that involve finger dexterity.

When the child begins to learn to write, the teacher may start by getting her to trace over and then copy the letters of the alphabet. In many cases this too will be slow and require a lot of repetition. Problems with forming the letters and spacing out words are common, but with practice progress will be made. Again, using letters and words to write sentences and stories on subjects the child is particularly interested in will aid her motivation.

Because children with WS tend to have co-ordination and motor problems, they may find it difficult to hold a pen or pencil properly, and drawing, copying and writing tend to be poorly

executed. The teacher may recommend that the child use a pencil grip -a device which fits on to a pencil or crayon and helps in pencil control. It will also prevent the child from grasping the pen/pencil too low down and thereby obscuring what she has just written. If your child is unable to exert sufficient pressure on a pencil or pen, you might find that giving the child thick felt-tip pens helps to overcome this problem.

Like most children, children with WS love working on computers and can practice useful exercises on them. If they can write and spell but have difficulty with pencil control, they could use computers for free writing, thus by-passing the difficulties of writing by hand. If you have a home computer, your child's teacher or the school's educational psychologist might be able to recommend and perhaps lend you suitable computer programmes for your child to work on at home.

The children's visual and motor problems may also mean that they are nervous of heights, and of activities such as climbing, going down stairs, walking along a bar in the gym etc. They may also find it difficult to throw and catch a ball, cut with scissors and ride a bike. All children will improve on these tasks with time, provided that they are given reassurance, support and plenty of practice.

Physiotherapists and occupational therapists can be very helpful in all of the above areas. They can assess and provide exercises for the child in the areas of co-ordination, balance, gross and fine motor activities, and they can advise on building up skills in dressing, washing, eating with a knife and fork and writing. You may wish to ask your child's Paediatrician, GP or health visitor about whether she could benefit from occupational therapy or physiotherapy, and request a referral.

d) Reading

The abilities of children with WS vary a lot, but many do learn to read at a basic or, in some cases, at a more advanced level. As already discussed, many of the children have relatively good verbal skills, including a very good memory for sounds and good auditory sequencing skills. In view of this, approaches to reading, which rely on these auditory skills, are likely to be most effective. The child should first be taught the names and sounds of the letters of the alphabet. This may well be a slow process requiring a lot of repetition and patience, but once the child knows the letter sounds, he can be taught to sound out the letters in a word and blend these sounds together to form the word. This is just one of a number of approaches that can be used to teach reading.

Where children are preoccupied with particular objects or topics (e.g. trains, pop stars, foreign places), introducing reading materials on these topics will enhance their interest and motivation to read.

e) Leaving School

The question of what will happen after the child leaves school can be a major worry for parents. Our findings indicate that most adults with WS continue to need at least some supervision and support in their daily lives. People with Williams Syndrome, like all of us, go on learning and acquiring skills after they leave school. They may go to a Further Education Unit or College, or other training scheme, for a number of years after leaving school, to continue their training in independence and self help skills such as dressing, cooking, independent travel, and to learn other skills. For the longer term, Special Education Centres, Adult Training Centres or Day Centres are further options, which provide work activities and recreational facilities. A few adults obtain employment either on the open market or in sheltered employment, and some go on to independent living. Others require ongoing support and supervision in everyday activities, and continue living at home, or move to residential hostels or group homes. Parents should try to obtain information about the available options well before the child leaves school. You can arrange to consult the school's educational psychologist or a special needs careers officer, and also

the local Learning Disabilities Team within Social Services about the options that are available, in anticipation of school leaving. Further advice can be found in two booklets available from the Williams Syndrome Foundation:

Udwin, O., Davies, M., Stinton, C. & Howlin, P. (2007)

Adults with Williams Syndrome: Guidelines for Families and Professionals.

Udwin, O., Davies, M., Stinton, C. & Howlin, P. (2007)

People with Williams Syndrome: Guidelines for Employers and Supervisors.

IMPLICATIONS FOR SIBLINGS

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 at times 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 other times they may feel guilty that they themselves are not disabled. 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 disabled sibling once their parents are no longer able to do so. Giving siblings the opportunity to talk about their feelings for their brother or sister with WS, and about their worries and anxieties, is important. Furthermore, realistic planning for the future can help alleviate many of their 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, or of developing the condition later in life. 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 its physical and behavioural characteristics. Siblings may also find it helpful to talk with the WS child'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 about the condition.

SOURCES OF HELP

a) Local Child Development Centres. These are organised differently in different districts but they tend to have multi-disciplinary teams consisting of paediatricians, psychologists, community nurses, speech therapists, occupational therapists and physiotherapists. One or more members of the team would be expected to assess and review your child's development at regular intervals. They can be a valuable source of help and advice about your child's development and any behavioural difficulties that may arise, and with your agreement can refer the child to other professionals if necessary.

b) Social Service Departments. These can provide advice on benefit entitlements. In many areas they can also organise respite care for children for a few days or weeks, for example at holiday times or during a crisis or emergency. Respite care can be helpful not only for giving parents much needed breaks, but also for enabling individuals to experience different environments, meet different people and gain independence skills.

c) Child Clinical Psychologists (employed within the National Health Service) and **Educational Psychologists** (employed by education authorities) are available to help with behavioural or educational difficulties.

d) Portage schemes and other Early Years programmes are available in many areas, providing trained workers who work with parents in order to teach their young children new skills and to help them develop in such areas as language and motor development, co-ordination, self help skills and socialisation. Information about such schemes, and also toy libraries, opportunity group and other facilities in your area can be obtained from your health visitor, Child Development Centre or Local Education Authority.

e) Benefits: Information about the benefits which you and you child are entitled to claim can be obtained from your local Social Services Department or by contacting Disability Alliance, Universal House, 88/94 Wentworth Street, London E17SA, Tel. 020 7247 8776 (www.disabilityalliance.org), or RADAR (Royal Association for Disability and Rehabilitation), 12 City Forum, 250 City Road, London EC1V 8AF, Tel. 020 7250 3222 (www.radar.org.uk).

USEFUL ADDRESSES AND FURTHER READING:

General:

Foundation for People with Learning Disabilities:

(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

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

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

Legal Provision:

Quin, 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

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

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

Family Fund

Unit 4, Alpha Court
Monks Cross Drive
Huntington
Yorkshire
YO32 9WW
Tel: 0845 1304 542
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.

ACKNOWLEDGEMENTS

We are grateful to Sir George and Lady Cooper and the Williams Syndrome Foundation for highlighting the need for information booklets on WS for parents and professionals. We would like to thank them, as well as Mike Adlam, John Nelson and the Foundation's past and current Management Committees for funding a series of research studies upon which this booklet has been based. We are grateful to Dr Neil Martin and other colleagues who read earlier drafts of the booklet and contributed helpful suggestions and comments. During the course of this research we have benefited from insights and information provided by many parents, teachers and children with WS themselves. We are grateful to them for all they have taught us about the condition.

Dr. Orlee Udwin Consultant Clinical Psychologist West London Mental Health Trust 405
Kennington Road London SE11 4QW

Professor William Yule Professor of Applied Child Psychology Institute of Psychiatry Kings
College London

Professor Patricia Howlin Professor of Clinical Child Psychology Kings College London