



GUIDELINES FOR THE CARE OF ADULTS WITH WILLIAMS SYNDROME

What is Williams Syndrome?

Williams Syndrome is a genetic condition that is evident from birth. It affects very many aspects of an individual's functioning throughout life.

Physical problems are numerous but among the most common are: cardiovascular and urinary tract abnormalities, hypertension, gastrointestinal complications, aching joints, mobility difficulties, fatigue, sensori-neural deafness, visual problems e.g. refractive errors (long or short sight, astigmatism), strabismus ('squint'- cross- eyes), difficulty judging distances and speed , and premature aging.

Behavioural difficulties include: lack of social inhibition/discrimination, attention deficits, fears phobias, generalized anxiety (often related to health and illness), preoccupations obsessions (e.g. certain types of machinery e.g. lawn mowers, washing machines) and perseverative behaviour (doing or saying the same thing over and over again).

Most individuals have an IQ in the intellectually disabled range together with specific difficulties in verbal understanding, numbers, planning, problem solving, everyday visuomotor and visuo-spatial skills (such as pouring a glass of water from a jug or going down stairs or escalators), and complex memory tasks. However, people with Williams Syndrome also have an unusual profile of abilities. This can result in many problems and misunderstandings. Thus, they tend to have a superficially good vocabulary and are keen to engage in "social chat" with others, including complete strangers. They often assume that complete strangers will feel friendly towards them and are shocked when they do not smile. This can lead to an assumption that they are much more able than is actually the case. In particular, verbal understanding is much less well developed than their spoken language would suggest. Similarly, their attempts to socialise frequently give a very misleading impression of their true level of social competence. In fact, social understanding is generally limited and adults with Williams Syndrome have a risk of getting into serious social difficulties if not adequately supported and supervised. Therefore, they are vulnerable to exploitation and abuse.

Why do people with Williams Syndrome experience many problems as adults?

Because of their very complex difficulties adults with Williams Syndrome are rarely able to live independently and continue to rely heavily on their families for support. Although they may seem very sociable they are rarely able to maintain friendships with individuals of their own age. Finding suitable accommodation is often challenging as adults with Williams syndrome may find it difficult to live in a close community. They may constantly complain about their environment, or the people who are living with or caring for them. They may make unwarranted accusations about being victimised or abused when there is no evidence of such behaviours. They may intrude into others' personal space or property and be 'over- friendly' compared to their peers. Some individuals can become very angry and resistant if their behaviour is challenged. Others become easily distressed by what they may incorrectly perceive as others' attempts to harm them, (e.g. if accidentally bumped into) or if they feel they are being ignored. There is a great need for attention and reassurance. These problems become more apparent if the adult is placed in an unsuitable environment or is under stress (e.g. if others have unrealistic expectations of their true level of ability).

Difficulties in communal living are also often exacerbated by oversensitivity to certain sudden noises (hyperacusis) such as vacuum cleaners and babies crying, which can be a significant

problem for some individuals with Williams Syndrome. Concerns and fears about physical illnesses are also common. However, it can be difficult to determine whether health concerns are indicative of over-anxiety and need for reassurance or whether they are actually due to physical problems. These can be relatively mild, such as joint pains or fatigue, or much more serious problems such as heart or kidney disease. Routine investigation of these complaints may be compromised by communication problems, anxiety over intrusive tests and/or professionals' lack of familiarity with the health conditions that are known to be associated with Williams Syndrome.

The combination of these difficulties quite frequently results in mental health problems such as anxiety and depression, which can become severe enough to warrant treatment.

Recommendations for helping adults with Williams Syndrome

One of the biggest difficulties for carers, doctors or other professionals is recognising that understanding in Williams syndrome is *almost always* well below the individual's level of spoken language and that the superficially good social skills disguise profound problems in social understanding and social competence.

Although all individuals are different the following general guidelines can be helpful in minimising problems.

- 1) Physical and mental health should be regularly monitored. For individuals who are very concerned about their physical health it is important to ensure that they have a full physical examination by a physician who is an expert in treating people with intellectual disabilities. Offering regular check-ups by a *named* individual can help to minimise uncertainty and avoid constant visits to other doctors or hospitals.
- 2) Behavioural and mental health problems are most commonly a result of social and environmental stresses but when these problems are present it is essential that the professional makes a holistic assessment of the individual's medical, social and environmental situation. In very many cases, appropriate changes to the environment can have a significant impact and should always be considered *before* medication is prescribed.
- 3) Use simple, concrete language at all times. Avoid complex and abstract explanations or concepts.
- 4) Remember that the person with Williams Syndrome usually understands less than may be apparent from the way they talk.
- 5) Provide a structured and consistent daily programme *throughout the year*. Breaks in daily routine (e.g. for holiday periods etc.) are often difficult for people with William's syndrome to cope with, and can result in increased behavioural disturbance.
- 6) Provide adequate support to reduce the risk of social vulnerability (especially when away from the home situation).
- 7) Ensure that all involved in the person's care agree on a *consistent* way of managing problems (such as how to respond to attention seeking; complaints against others etc.) This is usually best done by identifying ONE person to whom complaints can be made so that other staffs do not become engaged in these disputes. Rules should be clear, simple, and explicit.

- 8) Try to avoid giving attention for problem behaviours (such as complaints about others; constant seeking of reassurance). Make sure instead that the person achieves the security or attention he/she needs for other behaviours (e.g. helping in the home; NOT complaining about others; etc.). Concentrate on the “good” rather than the “bad”.
- 9) Reduce stress in the environment by eliminating excessive noise, unpredictable or unnecessary changes in routine or staff, or unrealistic expectations of what the individual is able to achieve/understand.
- 10) Try to build up daily life skills *gradually* and with adequate support in order to develop greater self-help and independence. This may require patience on the part of a familiar sympathetic caregiver and slow practice using a step-by-step routine for certain visuo-motor skills such as walking down stairs.

In summary, intervention should focus on the identification and modification of potential causes for behavioural difficulties *before* medication is considered. Medical and psychological issues should be investigated as part of the holistic assessment. If long-term medication is required for mental health problems this should only be prescribed by a doctor who is aware of the known associations of Williams Syndrome. If required, medication should rarely be given without assessment by a psychiatrist with expertise in the treatment of individuals with intellectual disabilities who in turn should be supported by local learning disability services.

Monitoring should include psychological testing (clinical and educational) by individuals familiar with learning disability and in particular, the Williams Syndrome behavioural profile. Many WS individuals have no regular assessment of their cognitive/intellectual abilities. Psychological therapies (CBT, mindfulness, etc or services available through the “local learning disability service”) should be investigated.

Other essential health checks for adults are documented on pages 12 and 13 of the “Guidelines for the Management of Williams Syndrome.”