GROWING UP WITH CdLS

CHANGES IN ADOLESCENCE AND YOUNG ADULTHOOD

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Introduction

FINDING ANSWERS TO GROWING QUESTIONS

What does the future hold for my child? What changes can I expect as my child grows up? These are questions which many parents often ask and, of course, there's never a definite answer.

When a child has a diagnosis of Cornelia de Lange Syndrome (CdLS), the uncertainty is likely to feel even greater. Many parents of people with CdLS have even had the horrendous experience of being told that their child may not live to adulthood.

While it is now known that people with CdLS can live well into their adult years, there has been relatively little information available on how the syndrome affects people as they approach and reach adulthood.

Research is now beginning to suggest that certain changes in behaviour and mood (and possibly other areas too) can occur as people with CdLS grow up.

Here at the University of Birmingham, Professor Chris Oliver, Dr Jo Moss and their team have also begun to see some themes emerging in personal accounts of late adolescence and early adulthood, particularly in people who are more mildly affected by CdLS.

We wanted to share with you some of the stories we've heard from families, to illustrate the different experiences of people with CdLS as they grow up.

In this article, we asked six young adults with CdLS and/or their families to tell us about their experiences of growing up with the syndrome (their stories are in Case Studies 1 to 6).

We try to show how parts of their stories might relate to the findings from research into CdLS.

Along the way, not only did we find out about some of the changes that these young adults and their families have noticed, but also about the inspirational ways in which they have coped with the challenges of adolescence and early adulthood. In the words of one parent, through all the difficulties, it is a "dignified, valid and beautiful journey".

When reading about the experiences of these young adults and their families, it is important to keep in mind that CdLS is extremely variable in its effects, both physically and psychologically. This is likely to be just as true of the changes people experience over time. Furthermore, disentangling "the CdLS" from all the other aspects of an individual is incredibly complex, and it is always difficult to say how much the findings from research on groups of people with CdLS relate to any one person. We can only describe the broad picture whilst knowing that everyone is different.

In the article, we've included some ideas from families for how to deal with certain difficulties. We also mention suggestions based on work conducted with other groups of people who experience similar problems (e.g., people with Autism Spectrum Disorders, as well as the typically developing population). More research is needed to find out how effective different strategies are for people with CdLS. The advice here is not intended to be anything like a complete review of possible ways to deal with the problems, but might provide a few pointers.

We've also put in some references to some of the papers whose research we mention. If you want to access these papers, or to get a summary of them, just send us an e-mail (our e-mail address is at the end of the article).

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What CHANGES?

Difficulty with Change and Unpredictability

Individuals with CdLS have a lot in common with those with Autism Spectrum Disorders (ASD) and, indeed, many people with CdLS also have diagnoses of ASD (See Box A).

People with ASD often strongly prefer routine and predictability, and it seems that this is also true of many people with CdLS. In the individual stories (Boxes 1 to 6), the families of Estey, Jake, Victoria, Keith and Eleanor all report change and unpredictability to be a major source of distress.

Families of people with CdLS sometimes report that problems with change and unpredictability increase over the course of adolescence. Some research also suggests that ASD-type characteristics might become more prominent over time, which could explain these increasing problems with change.

Data collected at the Cerebra Centre indicated that people

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Box A

Autism Spectrum Disorders

Autism Spectrum Disorders (ASD) are a diverse and complex set of developmental conditions. People are usually diagnosed with ASD on the basis of differences in:

- **Social interactions**
- **Communication (verbal and non-verbal)**
- **Repetitive behaviours**

Most people with a diagnosis of ASD do not have any known genetic syndrome. However, many people with CdLS have certain features of ASD, and some also have a formal ASD diagnosis (this is true of several other genetic syndromes too, such as Fragile X syndrome). This means that some theories of ASD, and some known approaches to dealing with difficulties, may also be relevant to people with CdLS.

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with CdLS over 16 years of age showed more repetitive behaviour and other ASD-like characteristics than did younger people with CdLS. Researchers in Italy (Basile et al, 2007) also found that repetitive behaviour was more common in older people with CdLS. Other research (e.g., Sarimski, 1997) suggests that older children (6 years and above) struggle more with changes to their routine than younger children.

One theory which is sometimes used to explain aspects of ASD is the "Executive Dysfunction" theory **(see Box B)**. This theory may also be useful to explain certain characteristics of people with CdLS.

The amount of change and unpredictability in life naturally increases around adolescence, and this could be one reason that people with CdLS have particular problems as they enter this phase of development. The transition from child to adult services very frequently poses difficulties (see especially Keith and Jake's stories – Case Studies 3 and 4).

Increased challenges in everyday living also accompany

Box B

Executive Dysfunction Theory of ASD

"Executive Function" is an umbrella term to describe a number of different abilities, including planning, controlling impulses, inhibiting highly practiced actions when they are not appropriate, and shifting to different thoughts or actions according to the situation. It is thought that some aspects of executive function may be impaired in Autism Spectrum Disorders. This is known as the "Executive Dysfunction" theory of ASD.

The Executive Dysfunction theory could help explain why switching from an established routine can be particularly difficult for people with ASD. This is because a change in routine might involve skills like inhibiting actions which people are used to performing, and planning and acting out a new set of actions. Because of the commonalities between ASD and CdLS, it could be that the Executive Dysfunction theory is helpful in understanding aspects of CdLS, too.

EXECUTIVE DYSFUNCTION AND ROUTINES

Imagine that a fictional person, Arlene, has in recent years always been picked up by a particular minibus driver, Bill, at 8.05am. Bill always knocks on the door and says "Good morning Arlene" when Arlene opens it. Arlene replies "Good morning Bill", and gets into the front of the minibus. One morning Bill is unwell and a different driver, Charlie, knocks on the door at 8.05am. When Arlene opens it, he says "hello, I'm Charlie" and offers his hand for Arlene to shake. When Arlene opens the door, she must suppress her previously learnt action of saying "Good morning Bill". She must also generate a new appropriate action (e.g., taking his hand, and saying "Hello Charlie"). Now imagine that Charlie opens the back door to the minibus rather than the front. Arlene must inhibit her usual action of getting in the front door, and potentially plan and carry out a whole series of other actions (either to explain the problem to Charlie or to get to a new seat on the minibus). To a person who has difficulties with Executive Function, all these things could be extremely difficult.

STORY ONE

Estey

Freedom presents its own challenges

Estey turns 31 this year, and lives in her own flat in Brighton. Her parents live just around the corner and see her regularly. She phones them if there's a problem (a blocked drain; too much noise from the neighbours). Estey also has a call button system and sensors in her bed due to her epilepsy, which started when she was 23. Estey pays for a cleaner to come every two weeks.

Last year Estey successfully completed a Work and Life Skills course at college. She is also an active member of her church, and helps out with Brownies and Guides. As well as CdLS (diagnosed only in 2009), Estey has a diagnosis of Asperger's Syndrome and socialises with her local ASD group, Aspire. A Christmas party with this group was where Estey met her boyfriend of over five years, whom she sees every Wednesday. Estey plays the drums, likes listening to music and story CDs (her favourite author is Roald Dahl) and watching films. Estey also has an interest in steam trains, and collects fans.

■ Issues in early adulthood

Estey's increased ability to live and go out independently – and to use public transport – has allowed her a lot of freedom, but has also presented big challenges.

In recent years, Estey has had problems when things don't go according to her expectations. For instance, she became quite distressed and angry when she was not allowed to get on her usual bus, leading to an incident in which the police became involved. It can be very upsetting for Estey when plans change unexpectedly (e.g., if a friend cannot make an arranged meeting), or if she has not been able to adequately prepare for a situation (e.g., if she hasn't brought the right clothes for an activity like cooking or walking). In these instances, Estey has had some problems with anger and aggression, as well as anxiety.

To help deal with these problems, Estey has taken part in Cognitive Behaviour Therapy (CBT). She has learned to walk away and listen to her music to cope with difficult emotions like anger. Estey also now carries a small information pack listing her diagnoses and difficulties, which she can show to people to help them respond to her more helpfully. It is not always immediately apparent to people that Estey has an intellectual disability, and that this might require special consideration.

Dealing with Estey's tendency to take things very literally has sometimes been tricky for her family and others. In the past, Estey has refused to speak to a policeman because she had been taught not to speak to strangers. Her parents have now explained to her that policemen (and certain other people in uniforms) do not come into this category. Estey's parents also take great care to explain exceptions to general rules like these, and to try to anticipate potential problems with instructions. Estey has a mobile phone, which she can use to phone her parents if there is ever a problem when she is out. This has been extremely useful to the family, as sometimes Estey's parents have been able to talk her through problems as they arise.

Estey's parents feel that her difficulties with anger might be related to the onset of her epilepsy, or to the medication she has to take to control it. They also feel that Estey has been a bit more sluggish in her movement in recent years and that her memory, and possibly her speech fluency, might have become slightly worse as she has taken the anti-seizure medication. In addition, Estey's self awareness has improved, and she has been more aware of her limitations. Her CBT therapist has helped Estey to focus on the things she can do, and to look at the positives in situations.

Over the years, Estey's parents have spent a lot of time advocating for their daughter, and have often had to fight to get the right support for her. They have found ventures to be much more successful when they have had good communication with the other people in Estey's life. For example, the Work and Life Skills course at college went well as a result of Estey's parents being in close contact with her teachers.

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enhanced independence (see especially Estey, Case Study 1). A good example of this is the use of public transport, whose intrinsic variability and unreliability has caused problems for many of the families we speak to. In Estey's case, an unexpected occasion when she was not allowed to stay on the bus in her usual routine led to severe anxiety and an aggressive outburst.

The degree to which increases in ASD-like tendencies might also be associated with neurological change over time is presently unclear, and more research is needed **(see Box C)**.

WHAT CAN WE DO?

Parents and carers deal with difficulties with change and the need for routine in different ways. Keeping life as predictable and regular as possible can be very effective (e.g., life at Jake's house; see Case Study 3). This might mean talking to schools, colleges and care staff to help them understand the need for routine. Preparation can also make a huge difference when something unusual is likely to happen. For instance, Keith's family finds that talking through the details of an unusual event with Keith before it happens can take the edge off the stress it causes. Transitions can be eased by allowing plenty of visits to a new place before a move occurs (like Jake's move from school to college, and Eleanor's move to her flat), allowing the process to occur gradually. Sometimes using Social Stories about moves and changes can help.

Estey and her family have found that use of her mobile phone (usually to call her parents) when unexpected events occur has been invaluable, as well as the Cognitive Behaviour Therapy (CBT) strategies she has learnt to use to cope with anxiety when changes happen. Allowing people with CdLS to organise certain events for themselves can be really helpful, as allowing the person some control might help to reduce any fear of unpredictability (e.g., Victoria's barbecue and, hopefully, her 30th birthday ball).

In some instances, not allowing a routine to develop in the first place can avoid some of the dependence on schedules which are often difficult to maintain. Jake's parents deliberately arrange their visits for different times from week-to-week so that there won't be a problem if they are ever unable make a specific day.

Box C

Neurological change

When we talk about "neurological" change in this context, we usually mean changes in the central nervous system. The central nervous system includes the brain and spinal cord.

Everyone's brain changes constantly over time, and in this sense we are all constantly undergoing neurological change. There are also specific neurological changes which tend to occur over time as people develop and grow up. It is possible that there are certain different or additional neurological changes as people with CdLS grow up, which might explain some of the possible changes we see in people's behaviour. However, further research would be needed to assess whether or not this is the case.

2

Mood, interest and PLEASURE

Research shows that age may play a part in emotions

Lisa Nelson, who was funded by the Foundation, recently completed her PhD at the Cerebra Centre with Chris Oliver and Jo Moss. She used a specific questionnaire (the "Mood, Interest and Pleasure Questionnaire" or MIPQ) to assess the mood of people with CdLS, as well as the interest and pleasure they show in activities.

Lisa found that there were some differences between the age groups. In particular, older people with CdLS displayed lower interest and pleasure than younger people. Other researchers, at The Cerebra Centre for Neurodevelopmental Disorders and elsewhere, have similarly reported that mood, interest and pleasure are lower in older children and adults with CdLS than in younger children (e.g., Berney, Ireland & Burn, 1999) and also suggest that adults with CdLS show higher levels of negative emotions than children (Oliver, Berg, Moss, Arron & Burbidge, 2011). Taken together, these studies might suggest that mood, interest and pleasure decrease as people get older.

The possible changes over time appeared to occur through

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Box D

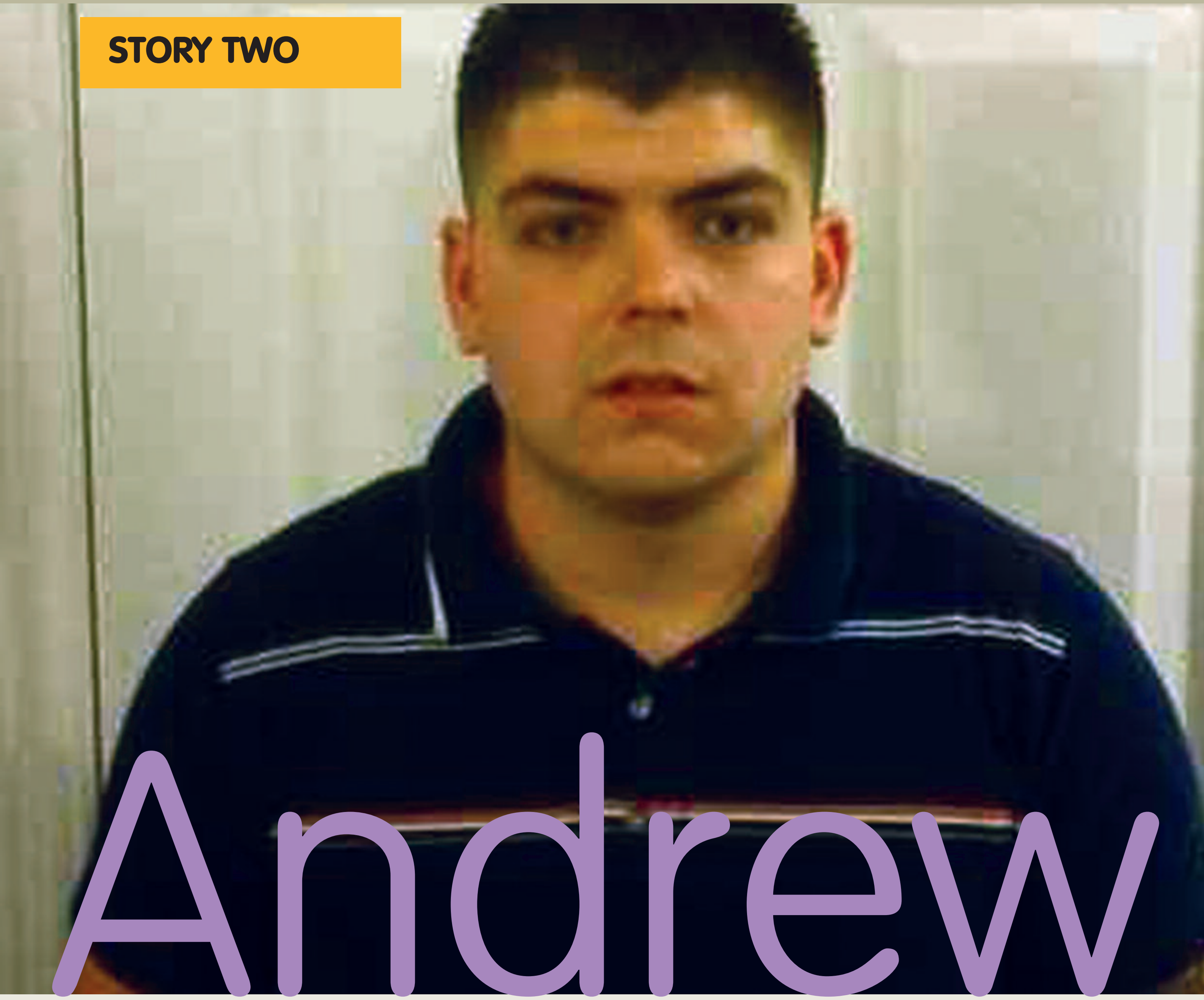
Changes with age

How do we know whether possible changes with age are general across all groups, specific to people with intellectual disabilities, or specific to CdLS?

This can be a difficult issue to tease apart, but often studies contain "comparison groups".

For instance, we might compare the behaviour of people with CdLS with those of people with different genetic syndromes, both with similar and dissimilar levels of ability. The possible developments with age of the some of the issues identified in this article seem to be relatively specific to those with CdLS.

STORY TWO



Andrew

Work options affected by issues with sleep

Andrew, 24, lives independently in a flat near the seaside. He has been living independently since 2006, and enjoys it. He lives close to extended family and sees his auntie and grandma from time to time. He speaks regularly on the telephone to his mum, who now lives in Scotland.

Andrew works two days a week at a local supermarket, a ten minute drive away. Andrew drives his own car, has an interest in motor vehicles and recently completed a diploma in Motor Vehicles and Repair. When we interviewed Andrew, he was awaiting his certificate and thinking about the future possibility of working in a garage.

■ Issues in early adulthood

Andrew says that his main difficulties are with sleep. He has always found it very difficult to get to sleep at night, and cannot fall asleep unless he has the television on. Then he has extreme difficulty waking up the next day. Sometimes he has slept through the noise of two mobile phones, seven alarm clocks, the TV on timer at full volume, and a lamp programmed to come on at the same time as alarms. This has limited Andrew's options at work, although he has now been able to arrange shift times which suit his sleep patterns. Andrew is due to meet with a sleep expert to discuss his difficulties.

Andrew feels that he is quite reserved and finds socialising difficult. Working in the retail industry, and particularly talking to customers, has helped with this, but meeting new people remains hard for Andrew.

Andrew has also had problems with low mood and anxiety. For instance, he has not been able to attend certain medical investigations (e.g., for a narrow oesophagus) due to anxiety about the procedures. However, Andrew has also been able to overcome some of his fears. He used to be afraid of needles, but he tackled the fear head-on by getting several tattoos (despite the fact that he initially passed out!).

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adolescence into early adulthood. There was some indication in Lisa Nelson's study that approximately 19 to 22 years of age may be a particularly low point for interest and pleasure. It is interesting that this is often the time people's living arrangements and day care change and there is evidence that disruptions to routine and an increase in predictability might be significantly related to low mood.

Several of the individual stories featured here are consistent with this lowering in mood, and decreased interest and pleasure in activities, in adolescence and early adulthood (e.g., see especially Estey, Andrew and Victoria; Boxes 1, 2, and 6). Some of the people we talked to also described how mood seemed to improve again after a period of difficulty, particularly when life became more predictable.

WHAT CAN WE DO?

The first thing is to be aware of possible signs that someone's mood may be deteriorating, as it might not always be obvious. Possible signs include changes in appetite, reduced motivation, loss of self help skills (e.g., washing, dressing), picking or scratching arms and legs and seeming tearful, tired or even physically unwell.

Second, if mood seems to deteriorate, it is important to consider whether this might be caused by pain or discomfort. Reflux, middle ear infections and tooth or sinus problems are all common causes of pain in CdLS. You and your doctor might also want to consider whether there is any evidence of kidney or bowel problems, which can be experienced by people with CdLS, and can affect people's mood. Your doctor should be able to help you to consider all these issues. Be persistent if necessary!

If you think there are ongoing problems with mood once any possible pain has been assessed and dealt with, then there are other things that can be tried to tackle the low mood directly. Ask your GP (or other healthcare worker) for a referral to Psychiatry and/or Clinical Psychology for assessment and intervention. There are various medications which could be considered (your GP or a psychiatrist might prescribe one of these). There are also interventions which do not involve medication, which clinical psychologists, some psychiatrists, and various other healthcare professionals should be able to advise you on and deliver. These can include Cognitive Behaviour Therapy, which helps people to improve their mood by changing what they do and/or how or what they think. Whether Cognitive Behaviour Therapy can be used depends on level of ability.

Finally, as discussed above, changes in routine and loss of familiar surroundings (like school) can affect mood, particularly in CdLS. It could be that gradually re-establishing routines will naturally lead to an improvement in mood over time.

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STORY THREE

Jake

Learning to deal with anger and anxiety

Jake (21) lives in a shared house with three other people with intellectual disabilities. A staff member is present 24 hours per day. He has lived there for two and a half years, and he, his family and the staff are thrilled with his progress.

Jake goes to college a couple of days each week. The rest of the time, he stays very busy and active with his housemates and the house staff, going for walks or to the shops, and playing football outdoors. He gets on particularly well with one of the other residents, Simon, with whom he recently went on holiday, accompanied by the managers of their house.

■ Issues in early adulthood

During his teenage years, while Jake was living with his parents, things got increasingly difficult. Jake was experiencing angry outbursts and at times became physically aggressive towards his mum.

There was also an occasion when Jake leapt out of the car and ran off while his mum was driving. The triggers for these episodes were often unexpected events, or changes to the usual routine.

Jake would become progressively more anxious (the external signs of which included pacing and picking at his skin), and then frequently the anxiety would turn to anger. Anxiety and anger about changes have always been an issue for Jake, but the problem got more extreme during his teenage years. With the help of a Trainee Clinical Psychologist, Jake learned some techniques to monitor and deal with his anxiety and anger.

Jake rarely has aggressive outbursts now he's settled into his new accommodation, but on one recent occasion he removed his bedroom door from its hinges and threw it in anger. After they had discussed the incident with Jake, it became clear to staff that he had been getting anxious about a particular upcoming event. Once he was able to opt out of this, things immediately improved. The staff at the house are learning how to spot earlier signs that Jake is becoming distressed, and to help him deal with anxiety by, for instance, playing a game of football outdoors. There are also certain members of staff to whom Jake feels he can talk when he is concerned about something. Jake has a great sense of humour, and certain anxiety-provoking events can be joked through with staff.

■ Transitions

The transition from school to college was very difficult, but ultimately successful. Importantly, Jake's school had a program which incorporates half days at college, and Jake was attending these with a school staff member every week for a year before he went. Some staff overlap between the school and college was also a great help.

Moving from home to his current accommodation was also hard for Jake and his family. The decision to make the move itself was difficult for them all, but they are now all extremely happy with the result. The family thought that a homely environment, living with other people, would suit Jake best, and chose his current house accordingly. Jake has now settled into the house brilliantly, much more quickly than anticipated. From around six months after moving in, his parents and the staff at the house considered that things were going quite smoothly, and the last year has been "fabulous".

Prior to the move, Jake's family worked though social stories with him about growing up and leaving home. Jake's parents (Myra and Steve) and Della, the manager of Jake's home, also all consider good communication between them to have been key to the successful transition. There was a 4-5 month assessment period prior to the move, during which Della and the family were in close contact, and this contact has been maintained ever since. The regular routines and the manager's assertive approach make Jake feel safe in the house. Visits from Jake's family are arranged in advance to keep things predictable and calm for him, and to avoid clashing with planned activities with the staff. However, as Jake settles in, the family increasingly feel that they can call any time, and at short notice, to ask staff whether they can pop in for a visit. For instance, Steve might pop by and take Jake out for an impromptu drink in the pub.

Clothing has frequently been a thorny issue for Jake. It has been important to him to wear specific things for specific occasions, and in the past he has often dressed like a policeman, or copied exactly the clothes which his father or grandfather wore. On a recent family holiday to the Lake District, Myra and Steve were surprised – and slightly apprehensive – when they noticed that Jake was not wearing his "going home" clothes on the morning they were due to return home. To be met by a casual "I'll change after breakfast" was to Myra and Steve a major indication of Jake's feeling relaxed and happy.

3

Anxiety, including SOCIAL ANXIETY

Triggers that lead to behaviour problems

Anxiety is generally defined as an unpleasant emotional state with feelings of apprehension, dread and uneasiness. It has physical aspects (e.g., racing heart) and psychological aspects (e.g., "worrying"). Anxiety can be very useful in certain situations (e.g., helping you to run away from a poisonous snake), but when it occurs too much or in situations when it is unhelpful, it can become a big problem. Anxiety is often closely related to low mood (see above), in that people who experience periods of problematic anxiety are also more prone to feeling "low" (and vice versa). The triggers for a period of anxiety vary widely from person to person. In CdLS, common triggers include changes in routine (see above) and demanding social situations.

All of the individuals whose stories feature in this article have had difficulties with anxiety during adolescence and early adulthood. This does not, of course, mean that everyone with CdLS will have similar problems (it should be noted that researchers in Italy found evidence that people who are more mildly affected by CdLS, like those interviewed for this article, may show more signs of anxiety than those with lower levels of ability; Basille et al, 2007). However, it does appear that certain types of anxiety might characterise CdLS, particularly as people get older (e.g., Kline et al, 2007).

Social Anxiety

It has been known for some time that social difficulties are common in individuals with CdLS. This is another area in which people with CdLS share characteristics with those with ASD. People with CdLS often experience anxiety related to social interaction (known as Social Anxiety). One extreme form of Social Anxiety, which is quite common in CdLS, is selective mutism. The term 'selective mutism' is used to describe the behaviour of people who will not speak at all in certain situations or to certain people, even though they are able to do so.

Research in our department indicates that social anxiety may increase as people with CdLS grow up (e.g., Collis et al, 2006). Social demands are likely to increase as people go through adolescence, and this might contribute to social anxiety problems (which are indeed common in adolescents generally). Further research might help us to under-

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STORY FOUR

Keith

Digital camera and laptop help get behaviour into focus

Keith is 24, and lives in rural Ireland with his mum, dad and four younger siblings. Keith's family found that the local day services on offer did not suit him, and he now has an individualised program of activities, aided by 12 hours of weekly support from staff who come to the house. The program of activities is carefully devised by his family and reviewed monthly by the support agency providing Keith's care. Activities are chosen based on observation of Keith's current skills, and on decisions about what skills would be most useful to gain to aid his independence. Keith's achievements are noted and the activity plan updated regularly. As part of this plan, Keith has attended eight separate days at his local college, and this has been a big success. He will attend eight more days from October 2011 to March 2012, when he will graduate with a Certificate in Contemporary Living.

When we spoke to Keith's mum, Catherine, Keith had the previous evening made dinner for his family. With support, he had bought and cooked the meat and prepared and cooked the vegetables.

Keith carries a digital camera around with him, and he and his family find this a wonderful aid to communication, helping Keith to express what he wants to say and providing an immediate basis on which to initiate conversation. Keith also saved for, and bought, his own laptop. He has recently participated in a local pilot program to make his own "digital biography", independently choosing the pictures and music, typing words with assistance, and including details such as which recipes he likes to cook.

Keith's family aims to provide him with as many opportunities for social interaction as possible, and social situations are also built into his activity program. Keith has built up a great rapport with people in the local community. He also has other friends with learning disabilities he has met through different activities such as art classes. Keith speaks to one of his friends on the phone every week, and she recently came to stay with him.

■ Issues in early adulthood

The period after Keith left school, and started at an adult day service, was extremely difficult for him and his family. When he was due to attend the day service, Keith would refuse to come out of his room in the morning, putting his bed across the door. The change in transport in itself proved tough – there were major problems with Keith getting to the day service on the bus, and Catherine describes how Keith would "freeze with fear" when he saw it coming. With hindsight, Catherine feels that Keith wasn't given the preparation he needed to cope with the changes that were coming up, and she feels that, in this respect, the system failed him.

Keith thrives on predictable routines and definite schedules. Unexpected events can cause big problems, with difficult behaviours such as holding tight to objects or people and not letting go when asked. Often associated with unexpected change, these behaviours also occur more generally in situations when Keith is frightened or asked to do something he doesn't want to do.

Keith's family feel that, when they realised there was nothing else on offer other than the day service, this was the start of a difficult and important journey; Catherine describes it as an initially very bleak time. A breakthrough for the family was when Keith had a Positive Behaviour Assessment, which helped identify triggers for Keith's anxiety, and some possible factors contributing to his holding behaviours. In addition, big improvements occurred when Keith was given his digital camera, and began to use it to interact so effectively. His laptop has also given him new ways to interact with people. Keith's family was initially worried about the class in which Keith enrolled to make his digital biography, because he did not know the 11 other participants, or the tutors. However, Keith appears to have become less socially anxious recently, and has thoroughly enjoyed the course; he and his family are extremely proud of the outcome. A further breakthrough was getting Keith his own car: care staff can now drive Keith to wherever they need to go without the stresses and unpredictability of public transport.

Keith would like, one day, to have the family garage converted so he could have it as his own home, and his parents are keen that this should happen in the future. Keith has also expressed an interest in working, and his family hopes that a job, perhaps doing some basic chores in a care environment, might become accessible to him.

This is not to say that Keith doesn't still experience problems, especially when things are unpredictable. There were difficulties over the summer when new and different staff were coming to work with him, and Keith's holding behaviours increased. However, his family has increasingly developed ways to minimise these (partly based on the outcomes of the Positive Behaviour Assessment). On next year's calendar, Catherine is already marking out the summer as a potentially difficult time for him, and plans to prepare Keith for this over the course of the year. She also feels that they are learning to negotiate the amount of information Keith needs in advance of events, in order to prepare him without causing him unnecessary anxiety in the lead-up. Catherine feels that over recent years, the progress Keith has made is extraordinary, and that 99% of the time he has a "dignified, valid and beautiful journey".

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stand whether there are also specific brain changes associated with increasing social anxiety in CdLS.

All the people with CdLS we interviewed for this article have shown signs of social anxiety as they have grown up. Andrew, for instance, often feels nervous in social situations and finds it difficult to get involved in conversation. Jake, Eleanor and Keith can get anxious when interacting with people, and have all had periods when they have shown selective mutism. However, it is certainly not all doom and gloom – their families feel that their social abilities have increased in recent years.

WHAT CAN WE DO?

Be aware of anxiety

The signs that someone is feeling anxious can vary, and may not be immediately obvious, especially in people who may not be able to easily tell you how they are feeling. However, they might include restlessness, pacing, forms of self-injury (mainly picking and scratching the arms and legs), irritability, loss of appetite or reporting feeling unwell.

Work on communication skills

Difficulty in making oneself understood is likely to worsen social anxiety. People with CdLS vary widely in their speech, and sometimes other communication aids might be useful. Visual aids can be a fantastic way to improve communication and the confidence to communicate. Keith's digital camera and laptop have revolutionised his social interactions (see Box 4). The National Autistic Society is likely to have many tips for helping people deal with social difficulties, which may also be relevant in CdLS.

Improving communication can also help clarify the causes of anxiety. It can be very easy to assume it is about one thing, when in fact it is about another **(see Box E)**. Clearing up any misunderstandings (sometimes due to over-literal interpretation or other communication problems) can reduce or even take away the source of anxiety (we do realise that this is easier said than done!).

Be aware of avoidance

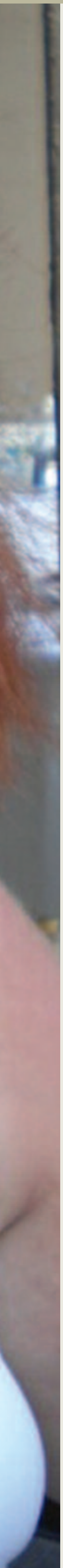
People (with and without intellectual disability) learn very quickly to avoid situations which make them feel anxious. It can be difficult for parents of people with CdLS to negotiate how much to go along with their children's decisions to avoid anxiety-provoking situations, and how much to persuade them otherwise (for instance, see Victoria's story, Box 6). We know that avoidance of a feared situation or object can make the anxiety much worse over time (because you never learn that you can cope with the anxiety, or that the situation/object is not really that frightening). On the other hand, increased control over what one does and does not do is a natural consequence of growing up, and it can clearly be important to respect people's own decisions.

Often people with CdLS are very keen to socialise, but still avoid social situations (perhaps because the anxiety is so unpleasant). Sometimes it is possible to gently encourage a person to socialise more, starting

Box E

Food for thought

Estey's parents have talked about a time when Estey was very distressed after a lunch event was cancelled, and they assumed it was the change of plan causing the distress. It later transpired that Estey was really concerned that she would not be getting any lunch.



STORY FIVE

Eleanor

Family support the foundation for independent living

Eleanor is 28 years old and lives in her own flat in Brentwood. With the help of her parents, Alison and Eddie, and 52.5 hours of support staff per week, she has lived independently for two years. Along with her support staff, Eleanor goes to yoga classes (sometimes she even takes her dad), swimming, the cinema, to get her hair and nails done, and to a weekly social group for people with intellectual disability. Eleanor also uses her computer and i-pod proficiently, and has become quite adept at solving technical problems herself (having tired of Eddie's self-professed i-pod phobia!).

After leaving school at 19, Eleanor moved to a college in Wales, where she lived in a variety of supported accommodation (always with staff sleeping in), on and off campus. After this, and prior to her current flat, Eleanor lived for five years at Fitzwalter House, a Mencap-run home whose purpose is to prepare people for independent living.

■ Issues in early adulthood

Because Eleanor has lived in several places over the course of her adult years, she has had to deal with a number of transitions. Eleanor takes a long time to get used to people, being initially very shy in their company, so moving between different sets of staff has been tricky. The success of her most recent move – to her current flat, and to independent living – was dependent on several factors. Eleanor had a good relationship with some of the staff at her previous accommodation (Fitzwalter House). A particular support worker (Paulo), with whom Eleanor had great rapport, continued to work with Eleanor for a period after her move to Brentwood. During this time, staff from the local support agency shadowed Paulo.

Then, when Eleanor seemed ready, Paulo left and the new staff took over. Eleanor's parents also slept in the flat with Eleanor for the first two weeks, and sometimes also stayed during the time before and after the carers were in during the day. Eleanor has a great memory for visual symbols and pictures, and her flat is equipped with numerous visual aids. For example, there are pictures on the fridge to help Eleanor prepare food, and pictures of Eleanor's parents on speed dial on the phone (Eleanor has also now learned to use the phone memory to call her grandmother). Eleanor often calls her parents several times a day, sometimes for a chat and sometimes to sort out a practical issue. Eleanor's mum and dad feel that the success of Eleanor's independent living has depended heavily on their ability and willingness to put in a lot of effort and time to support her. They communicate regularly with carers, both face-to-face and through a diary kept in Eleanor's flat, in which they and the carers can write messages and reminders for each other.

Eleanor has always had a strong preference for routine and a dislike of change. Her parents feel that this may have become more extreme in recent years, and that Eleanor is a bit less easy-going than she was in the past. Since Eleanor reached her 20s, she has tended to get more anxious about things. She will ask the same questions repeatedly in the approach to events, seeking reassurance that things will be OK, and clarity about exactly what will happen when. Anxiety can also result from Eleanor's literal interpretation of language, or when she just focuses on specific words in a phrase. Recently, she became quite distressed that a favourite member of staff was "leaving". It later transpired that she had in fact been told that this member of staff was going on "Annual Leave". Sorting out the misunderstanding ended Eleanor's anxiety.

Over the past two years, there have been times when Eleanor has become aggressive, throwing things around her flat and sometimes causing breakages. As a result, she and her parents have decided that it is no longer sensible for her to have the heavy glass coasters she used to have, so these have been taken from the flat. Eleanor has also begun to self-injure, at times biting her hand and hitting herself in the face. Episodes of aggression or self-injury usually follow from anxiety. Eleanor's dad also feels that having to wait (e.g. when she wants an answer to a question) can trigger these behaviours.

Despite all the difficulties and changes in her adult life, Eleanor is thoroughly enjoying living in her flat. Given time and practice, she becomes comfortable with new routines. The first time she was expected to walk through scanners and have her baggage searched at airport security, Eleanor was very upset and became angry with the security guards. However, she has subsequently learned the routine and now it causes no problems at all. Eleanor has also become more agile recently, since losing a lot of weight at Weight Watchers (where she goes with one of her carers). Despite difficulties getting the words out when she is upset, Eleanor's speech also seems to have got better recently, and she is more willing to try to say more difficult things. She is also now brilliant at speaking on the phone. It has taken – and continues to take – a lot of hard work by Eleanor's family, but they generally feel that moving to more independent living has been very good for her.

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in relatively easy situations and working up to situations which cause more anxiety. Periods of anxiety are generally self-limiting, and sometimes it is a question of helping a person to tolerate some anxiety at the beginning of a social situation in order to learn that the anxiety passes and the situation becomes enjoyable. We need further research to establish the best ways to deal with social anxiety specifically in CdLS.

Getting help

As with other mood problems, if the difficulties are severe your GP might suggest a referral to a psychiatrist or psychologist.

Challenging behaviours

Researchers in Italy (Basile et al, 2007) studied 56 people with CdLS, between the ages of 1 and 31. Challenging behaviour, self-injurious behaviour and repetitive behaviour were more common in older people. Researchers in the USA (Kline et al, 2007) also found that the onset and development of behaviour problems was frequently a main concern for parents and carers of adults with CdLS. Several families have told us of an increase in certain types of problem behaviour over adolescence and early adulthood. In particular, angry and aggressive outbursts and self injury **(see Box F)** can increase, or even appear for the first time.

Estey's, Jake's and Eleanor's stories all contain examples of this increase in certain challenging behaviours.

We can't be sure what underlies this. However, there are some possibilities. First, anxiety frequently precedes angry and aggressive outbursts. We have seen above that there may be increased anxiety with age, perhaps partly related to increased difficulty in coping with unpredictability and changes in routine.

Certain other changes in the ways people think and act may also relate to these problems. For instance, research in our team (Oliver, Berg, Moss, Arron & Burbidge, 2011) suggests that adults with CdLS may experience more "impulsivity" than children (under 18). "Impulsivity" is the tendency to initiate action without forethought, which could contribute to some of the problem behaviours people describe (see, e.g., Victoria and Estey's stories).

WHAT CAN WE DO?

It doesn't need stating that self injury and aggressive behaviour can be extremely distressing and difficult for parents and carers to deal with.

As before, the first thing to check is whether pain or discomfort could be contributing to any change in behaviour. Then, it may help to address any problems with low mood and anxiety, try to keep routines predictable, and keep communication as effective as possible. Try to work out the situations which trigger the behaviour, and whether they occur only in specific places or with specific people. Keeping a diary of these outbursts and the events which preceded and followed the behaviour may be helpful.

Finally, bear in mind that a challenging behaviour may become "functional" socially, even if it does not start out that way **(see Box G)**.

Box F

Aggressive Behaviours and Self Injury

"Aggressive" behaviours are any behaviours which have the potential to cause injury to another person. These can include hitting, kicking, throwing objects, etc. When we use this term, there is no implication that the person necessarily intends to cause injury to anyone.

A person's behaviour is said to be "self-injurious" if it has the potential to cause harm to the person carrying out the behaviour. This can include when a person bites or hits themselves, or bangs their head on something. Again, there is no implication that the person necessarily intends to harm themselves.

N.B. Overall, self injury, whilst occurring in two in every three people, is no more common in CdLS than would be expected for the level of Intellectual Disability. Aggression is significantly *less* common in CdLS than in people without CdLS with a similar level of ability. It is the increase over time which the research points to.

Box G

Breaking the patterns

Imagine that a fictional person, Bob, bites his hand in response to feeling pain. When he does this, suppose that carers ask "what's wrong? Do you want to go out and play cricket?", because Bob is a person who loves to play cricket, and carers naturally want to stop him from injuring himself.

Now suppose that Bob later wants to play cricket, but his carers are too busy. If Bob asks "can I go outside?" he will get the answer "no". However, if he remembers what happened earlier, and bites his hand again, he may well be taken to play cricket to stop him from hurting himself.

The behaviour could very quickly become the most effective way of getting a game of cricket. It can be very difficult to avoid these patterns from building up, because the priority is to keep the person, and others, safe by stopping the behaviour as quickly as possible.

The problem is that the quickest way to stop a behaviour is also often quite rewarding for the person showing it. These patterns can be difficult to break once they're established – but it is possible.

If behaviours have become severe or persistent, it might be helpful to ask for a referral to someone who specialises in Challenging Behaviour, such as a clinical psychologist working in the area. If you would like a booklet and DVD on managing challenging behaviour, please send us an e-mail (our e-mail address is at the end of the article).

STORY SIX

Victoria

Working around a fear of glass

Victoria is 26 and lives in a flat in Essex, a short drive from her mum and dad, Jane and Alan. A member of staff is present in the building's office 24 hours per day, to support Victoria and the residents of the other flats. Victoria does not frequently need help from the support staff, although she sometimes goes to chat to them if she feels lonely. Victoria's mum , Jane, comes round at least every week for a social visit, and to help with household tasks.

When she was 19, Victoria moved to Grimsby to spend three years at college. She didn't initially want to go, but when she arrived she had a great time – and by the end she didn't want to leave. On returning from Grimsby, Victoria's social worker facilitated her move to her current block of flats. She eased the transition by visiting Victoria and meeting with her family in Grimsby before the move.

■ Issues in early adulthood

Over the past five years or so, Victoria and her family have faced some difficult issues. Victoria has developed a strong fear of glass and crockery, following an incident in which she swept some glasses off a table during an angry outburst in a restaurant. It can now be difficult to persuade Victoria to go out when she suspects there may be glass or crockery present. It is unclear whether Victoria is fearful that she might smash glasses/crockery (as in the initial incident), or whether she has developed a phobia of these items.

Although it can be very difficult, Victoria and her family work around this problem. Victoria has only plastic crockery in her house. Also, her family have at times been very successful in persuading Victoria to come out even when she doesn't want to, and have found that she then enjoys herself.

However, there have been numerous occasions when Victoria has not attended events which her family feels she would have enjoyed. Victoria's interests appear to change quite quickly, and she will sometimes stop doing some activity without warning or explanation.

Having control over plans helps Victoria. For instance, she has been able to organise and attend a barbecue in her block of flats, and enjoyed being able to control aspects of the event. Victoria's family think that a recent prescription of antidepressants may also be helping with Victoria's anxiety.

Victoria is very active on social networking media. She has a fantastic sense of humour and can be very sociable in the right context. When I met her, Victoria was looking forward to flying to France for a holiday – with her dad, Alan, as pilot. Victoria is also already planning and saving up for her 30th birthday party, for which she wants to have a ball.

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Other possible CHANGES

Movement, sleep, speech and causes

Movement

We have had some reports, and there are some reports in the research literature, of young adults with CdLS experiencing changes in their movements over time. Movements may become slower and more difficult to initiate. However, there has been very little research on this, and what we know is mostly limited to individual reports. We know many more young people with CdLS who have not reported these changes. We have also spoken to a number of parents who say their son or daughter's physical abilities have generally improved over time (including Jake and Eleanor).

Sleep

Some studies have found that over 50% of people with CdLS have some sort of difficulty with sleep (such as trouble going to sleep or staying asleep, or daytime sleepiness), although others estimate figures as low as 12%.

There has been some indication (Kline et al, 2007) that, in some people, sleep problems may get more severe with age.

One of the people we spoke to for this article (Andrew), cites sleep as his biggest difficulty. Andrew remembers having sleep problems even as a child, but they may have got worse since adolescence. Also, they can be more problematic now that he is holding down a job.

Sleep problems can be associated with reflux, so it is important to get this checked. Also, you might ask your doctor about checking for other physical conditions which can affect sleep, such as obstructive sleep apnoea. There is some evidence that people with CdLS have a higher than average rate of sleep apnoea. This is characterised by snoring, pauses in breathing, and night-time gasping and choking. Measures to deal with anxiety and/or low mood could also be important.

There are also plenty of books and websites for setting up routines for a good night's sleep. However, if these things don't work, your child's GP (or other doctor) might suggest referral to a sleep expert, or trying a prescribed sleep medication.

Speech

Some preliminary research within our team suggests that certain speech problems are more common in older people with CdLS, particularly the ability to "get words out". This work is at an early stage. We have also heard several stories of speech and communication getting much better as people get older.

Relationship between the changes

In this article, we've discussed several possible types of change which might occur as people with CdLS grow up. It may be that some of these changes are highly related to one another. For instance, an increase in ASD-like tendencies (e.g. problems with changes to routine) might contribute significantly to increased problems with anxiety or low mood. Further research might help us to be more clear about whether some changes "cause" other changes.

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CONCLUSIONS

Research into the long-term development of people with CdLS is still at a relatively early stage, and many questions remain.

What is the exact nature of any changes with age, and how specific are they to CdLS? How many people experience these changes, and in what ways? What is the cause of the changes? How are they related to each other? What are the different contributions of changes in the environment and changes in the way the brain (or other body part) functions?

We are pleased to say that the CdLS Foundation has continued to support this research by funding Lisa Cochran's PhD and she is currently following up people with CdLS, to describe any changes that have taken place. The Foundation has also agreed to fund a new PhD student, Victoria Johnson, who has just started working on how we assess change and what might be helpful for people who are experiencing change.

Adolescence and early adulthood are turbulent times for everyone. It may feel daunting (or even frightening) to read about some of the particular problems which can occur in CdLS, especially if you have a young child who has the syndrome. It is very unlikely, however, that any one person will experience all of the difficulties discussed in this article. Human development is such a complicated process, and much of the research is at an early stage. And, whatever changes do occur, and however they might manifest for an individual person, there are plenty of things which you and other people can do to help your child. After particularly difficult phases, families often readjust and find arrangements which allow things to get easier again.

It's been fantastic to meet young people with CdLS in researching this article, and to speak with their families. Very many thanks to everyone for taking the time to speak to us. Thank you also to all the other families who take part in our research at the University of Birmingham. We look forward to seeing you all again.

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