

Information for
parents who are
considering whether to
talk about an extra X
chromosome with
their daughter and
family members.

A Guide for Parents

Introduction

Each cell in our bodies contains structures called chromosomes. Chromosomes are bundles of DNA that contain a 'blueprint' for building our bodies. Two chromosomes called X and Y differ in men and women; males usually have one X and one Y whereas females usually have two Xs. The X and Y chromosomes are commonly known as the sex chromosomes. Sex chromosome trisomies occur when there is an extra copy of one of these sex chromosomes. The term 'karyotype' is sometimes used to refer to the number of chromosomes an individual has. Girls with an extra copy of the X chromosome have the karyotype 47,XXX, also known as Trisomy X or Triple X Syndrome. Trisomy X is found in around 1 in 1000 girls. The 2011 Census shows that there are around 32 million females living in the UK; this means that there are over 32,000 girls and women in the UK with Trisomy X. Many of these cases go undiagnosed, because the impact of the extra X chromosome is usually quite mild, with minimal or no symptoms.

Trisomy X may be discovered when a mother undergoes prenatal screening (amniocentesis or CVS), or after birth if a child has their chromosomes tested as part of a medical investigation. When a sex chromosome trisomy is discovered, parents have to decide whether to tell their daughter about this, and if so how to go about this. We found that this was an issue that concerned many parents but very little was known about how parents decide to tell their child or the process through which they do this. Many parents were also concerned about whether to tell family members, or others such as teachers.

As part of a research project at Oxford University, we held a Study Day on Trisomy X in collaboration with Unique, the rare chromosome disorder support group, during 2012. Some of the parents who attended agreed to take part in a series of focus groups. During these

focus groups we discussed how parents decided whether to tell their daughter about having Trisomy X. For those who did tell their daughter, we explored the different strategies that they found useful in disclosing to their daughter, and also to other children or family members. In addition, we had a focus group for teenagers and young adults with Trisomy X, and we also obtained information from other individuals who agreed to be interviewed by phone or by questionnaire.

It is clear that there are no right or wrong answers to the question “Should I tell my daughter about Trisomy X and if so, how?” Different families have adopted different approaches, and the decisions they make will depend on personal circumstances. Nevertheless, we think it is helpful to describe a range of factors that need to be considered in making this decision. We hope that the information here will be useful for parents who are considering whether and how to disclose about Trisomy X to their child, and to other family members and associated professionals.

One important note of caution: It is impossible to describe a ‘typical’ child with Trisomy X, because they are so variable. In general, parents who are not worried about their daughter’s progress are less likely than other parents to be involved in support groups or get involved with research. This means that much of the information from our focus groups comes from families whose daughter is experiencing some problems. It is likely, too, that these parents will feel more reason to tell their daughter about Trisomy X, as this can help explain developmental difficulties. We hope that parents reading this booklet will get an impression of the range of issues surrounding disclosure of a diagnosis, and may be able to relate to some of the examples we give of people’s experiences, but it should not be assumed that what is right for someone else will necessarily be right for your family.

Disclosure of a diagnosis

Most parents who have a daughter with Trisomy X will, at some point, think about telling their child about their genetic condition, and this may be something that you are currently thinking about. When we started working in this area, we found this issue was a source of concern to many parents, but there was little information or advice available to help parents like you in making these decisions.

The main questions that you will need to consider and that we will discuss in this guide are:

- Whether to tell your child.
- If so, how and when to tell your child.
- Whether to tell other people, such as family members or teachers.

Should I tell my child?

Parents typically face a dilemma in deciding whether or not to disclose a Trisomy X diagnosis to their daughter. On the one hand, many argue that they feel the need to protect their child from any harm, stigma or undue stress that might be caused by their knowledge of their chromosomal condition; on the other hand, many families also feel that it is important to be open and be able to provide their child with information that may be relevant for future decisions, for instance about schooling.

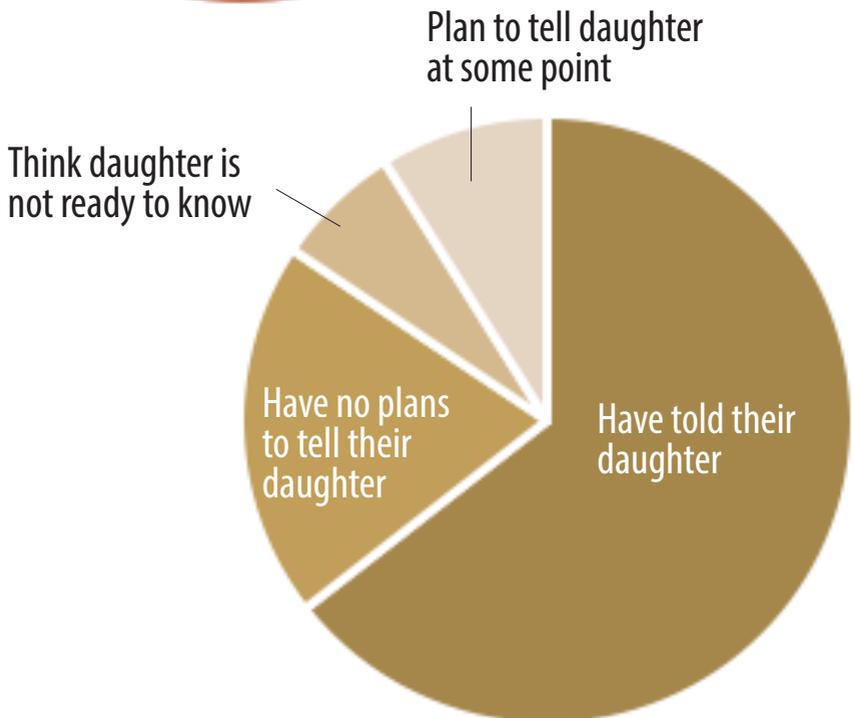
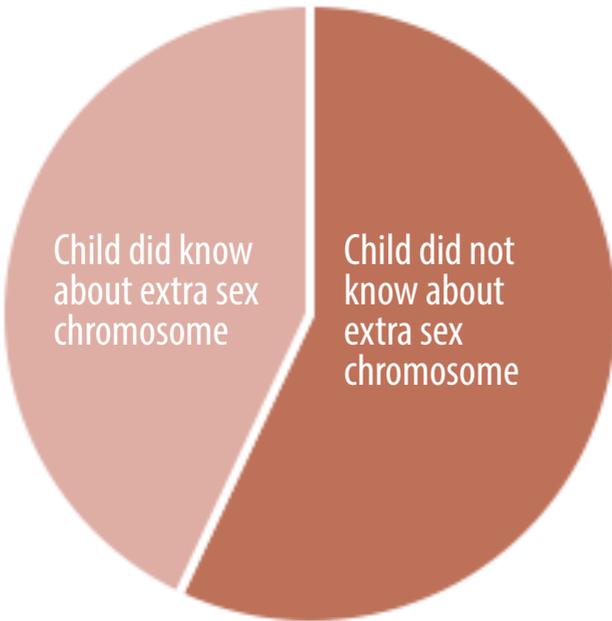
Protect child from stigma, harm or undue stress.



Be open and give them information that may be important.

In 2008 we carried out a study with parents who had a son or daughter with an extra sex chromosome. Of the 126 families who took part in our study, just under half of them (54 families) had told their child about their trisomy. In a more recent study with fewer families, we found a slightly higher number, 29 families out of 45 had told their daughter about Trisomy X. Out of the 16 families who had not told, 9 families had no plans to tell their daughter about her Trisomy X, and others planned to do so when she was older.

We summarise here the experiences and comments of parents who have a daughter with Trisomy X. We hope they will help you to weigh up the pros and cons and come to an informed decision about whether or not to tell your daughter about her extra sex chromosome.



Insights from parents discussing reasons against telling your child

Disclosure may stir up problems and do more harm than good

As far as we know, having Trisomy X does not affect a person's ability to have children, and there is a very low risk of Trisomy X being passed on to children. The impact of having Trisomy X on children's development is incredibly variable. Some girls have no obvious difficulties, whereas others may have problems affecting physical development or educational progress. If your daughter is progressing well, then you might think that disclosure would not be worthwhile.

Some parents may decide that telling their daughter about Trisomy X will only create needless upset and may leave her feeling stigmatised. As one parent explained, "It is, it's a label. And that's what I'm scared of – I don't want her to be labelled."

Karen explains how her youngest daughter Caroline is doing as well as her elder brother Michael despite having Trisomy X. Karen hasn't told 9-year-old Caroline about having Trisomy X as she's currently doing fine; both developmentally and at school:

“ Michael did everything really, really early – we didn't have any issues. Caroline was probably a bit later. When we saw the last genetics consultant he said, in his opinion, she's actually advanced.

With Caroline I just think 'I hope she's going to be okay'. It's that 50/50 – will she/won't she? At the moment, she's doing fantastic. If we didn't know about her XXX, I wouldn't be thinking twice about 'is she different' – I'd be thinking 'no, she's exactly like her brother was'. So if anyone ever said to me 'your child has got XXX now', I'd probably be like 'no, you're telling me the wrong information, she's fine.' ”

Concern that your daughter will use Trisomy X as an excuse

Even if your child is experiencing problems, you may still feel it is better not to disclose to her about her chromosome status. Several parents in our focus group expressed concern that awareness of a trisomy might provide their child with an excuse for behaving badly or simply not trying. Julia explains how she has been considering telling her 9-year-old daughter, Zoe, but the thought of Zoe using her Trisomy X as an excuse to not try is making her question the decision:

“ I’m at that point where I think I better do it now before the hormones really hit in... not quite sure. But I think, yes, it’s a hard call – do you tell or don’t you? Because really, she could go straight through and she’s no different from any of her other friends. But it’s also, I don’t want her to make excuses for herself – ‘oh I can’t do that because I’ve got XXX’. You don’t want that – you want her to be able to say ‘no, actually I can – I can try’. And that’s where it’s more difficult to balance.”

Lack of information or understanding

Trisomy X can be hard to understand even if someone has a background in biology. Some parents may feel it is counterproductive to tell their daughter that they have a genetic condition, if they can’t explain what it means or answer questions about it. The fact that there is wide variation from child to child just makes matters worse: telling your daughter that she has Trisomy X doesn’t allow you to say for certain whether or not she’ll have problems at school or home, and may just cause your daughter to feel anxious. – As a parent, it may leave you feeling inadequate when trying to talk about the diagnosis.

Imogen describes how, when receiving the initial diagnosis, the information provided by the geneticist was too complex and, although she decided to tell her daughter, Imogen explains that her own understanding of the condition has led to her daughter, Natalie, not fully understanding about Trisomy X:

“ And she [the geneticist] tried to explain the best she could, but it was out of our league really for what we understood. Well, it’s all above our heads really...once she started talking to us about the genes and chromosomes and that. Didn’t really understand it, did we, at all? But I think she tried her best with us at the time. I don’t know what would have helped, really. We didn’t leave with any information and it would have been nice to have had something to go home and go, ‘Ah!’ and really read it.”

On whether her daughter understood when she was told: “No, of course not, but then we didn’t either when we were told. But she knew that there was an X that came from mum and an X from dad, and she knew that she had an extra one and

we are still learning ourselves about what it is. So of course she didn't understand fully, no. But she has always known. Coming to these [Unique XXX events] with her has helped us all really."

Imogen's experience is interesting as, although she felt that her own understanding of Trisomy X wasn't complete, she still decided to tell her daughter and believes that attending events on Trisomy X has helped her whole family learn more about the condition together.

Feelings of guilt

A final consideration that might come up when deciding whether or not to disclose is related to guilt. Parents may be reluctant to open up a topic that they find incredibly stressful. One parent, Karen, spoke of her guilt at having a daughter who she knew had Trisomy X. She had refused a termination of the pregnancy, but had no idea how Trisomy X would affect Caroline's life:

“ Having to decide [about a termination of pregnancy] on a child who's going to go 50/50, one way or the other – I felt like I was playing God with her life and I still do because I think 'have I made the wrong decision in letting her live' ... did I make the right choice? Because I don't know what's in store for her and life is hard enough and have I given her a disadvantage because it's going to get tougher and tougher and people are not nice generally. I just think it's hard and have I taken the opportunity away from her or have I given her an opportunity? I just don't know how to see that at the moment and I still hold a lot of that guilt.”

Whilst Karen's feelings of guilt might not be uncommon among other parents who have a daughter with Trisomy X, it is very clear that as a parent you are not to blame if your daughter is born with this condition. Sex chromosome trisomies can happen to anyone and accepting that you had no part in this is incredibly important and should not stand in the way of any decision to disclosure to your daughter.

Insights from parents discussing reasons to tell your child

So far, we have considered reasons why parents may decide against telling their daughter about Trisomy X. We turn now to look at the question from the opposite viewpoint: that of parents who felt it has been beneficial to tell their child.

An explanation for difficulties

If your daughter is experiencing difficulties at school or with her behaviour or emotions, then telling her can be positive in enabling her to understand why she might be struggling in comparison with her friends. Children may find a diagnosis empowering, as it allows them to feel that any difficulties they are experiencing are not their fault, and that they can make informed decisions based on solid facts.

Pam explains that there came a point when her daughter Alexandra, started asking why she wasn't achieving as well as her friends and Pam sought out the help of a genetics consultant to help her tell her daughter about Trisomy X:

“ There was a critical point in her understanding when she was about aged 13 or 14 and she was beginning to notice a real difference in her educational achievement and started to ask questions. I was keen to meet with the consultant and my daughter, which was really helpful, and the consultant gave my daughter her phone number and lots of information and said to call whenever she wanted any more information or had any questions.”

In Pam's case, Alexandra didn't start to question a difference between her and her friends until later in her education, whereas Imogen told her daughter Natalie, very early on as she was already noticing that Natalie was falling behind at school:

“ I thought she should know really soon after she started school because..., as soon as she could. Yeah, they want to know why they can't keep up with the others. They have got learning difficulties, or Natalie has and I just felt that she should know why she couldn't keep up.”

Both Pam and Imogen's experiences indicate that they felt that their daughters knew that they were not learning at the same rate or progressing as well as their peers and that disclosing to them about having Trisomy X was providing them with an explanation for that. Imogen's daughter, Natalie, explains that being told about having Trisomy X gave her an explanation for the difficulties she was experiencing at school:

“ My mum told me when I was little so I have just been brought up with it. I think that’s the best way to be honest because at school I was always behind and I didn’t want to like, if I didn’t know I had this XXX thing like my learning difficulties and stuff, I would feel really weird and like not understand why. As soon as I found out I was like ‘oh yes’, like at school I can just be like ‘oh it’s because of XXX; I can put it as a reason.

I think I would feel different [if I didn’t know] then because then I would feel like I am out of place that I have got all these things that are a little bit hard for me. So I think it is best I would rather know because then I feel more myself. It’s good to have a reason.”

Another parent, Sarah, describes her struggle in deciding whether or not to tell her daughter, Olivia. Although Olivia isn't struggling at school, Sarah discusses her desire to tell her daughter about Trisomy X and points to it being a helpful explanation for Olivia's emotional difficulties:

“ She’s nine, so it’s imminent for us, but we’ve been holding off because we were so worried I’d just confuse her because she does get easily confused. But I do think we are approaching the time when I would like her to know. I don’t know why, I just think she does need to know. Because although academically she’s doing okay, she’s middle of the road, she’s not struggling but she’s not way ahead, she just gets on with her work and she copes okay. But I would say emotionally she suffers and I’d like to explain to her that that might be why”.

Avoidance of secrecy

If your daughter is not demonstrating any noticeable problems then, as mentioned previously, you may feel this is a compelling reason to not tell them. However, it is important to consider that they may still appreciate being told and indeed may find out at some point in the future. In families where a child has found out later in their teenage years or in early adulthood, there is a possibility of resentment or disappointment at not being told.

Anne's daughter, Abby, explains that if her parents had kept the news from her until she was an adult, she would have been disappointed:

“ Being 30 and not knowing about it, you would just be like, ‘why didn’t I know about it?’ That they [your parents] kept something that was, well yes wrong with you, well not wrong with you like literally wrong with you, but yes, that they have kept something from you and all this time you have had like problems in school and everything and you just thought it was a wrong reason, but then you knew it was XXX.”

Rebecca, who has Trisomy X, has just finished University and describes how she felt when she found out about having Trisomy X at 16 and how she dealt with her feelings that her parents had kept the information from her despite her struggles at school:

“ When I arrived at University I’d only known about my XXX for 2 years – my parents only informed me of it after abysmal GCSE results. I was shocked when I found out, I remember going through a period of not talking to my parents, but at the end of the day when is the right time to tell your child they have something like this? For me, it honestly just made sense, I’d struggled in school and had believed peers when they’d teased and said I was stupid.”

Rebecca's experience, in particular, demonstrates that although she understands how difficult her parents found the decision about when to tell her, finding out after many years of struggling at school was incredibly difficult for her.

Previous research on family communication about inherited genetic disorders has found that parents generally feel a responsibility to tell their children and provide them with adequate information about their condition¹. Although we have emphasised that there are no major consequences of Trisomy X for most girls, very little is known about Trisomy X in adulthood. There are many cases of women with Trisomy X who have had children, and the children typically don't have an extra chromosome. However, there are occasional medical reports of women with Trisomy X who have an early menopause. This could just be a chance association; it's not yet clear whether this is more common in women with Trisomy X than in other women. We hope that as more research is done, the situation will become clearer. One reason in favour of disclosing the diagnosis is that, as we learn more, the medical advice regarding Trisomy X may change – for instance women with this condition may be advised not to delay starting a family. If your daughter is aware of her diagnosis, it will be easier for her to discuss such matters with her family doctor, so she can plan appropriately.

¹ Metcalfe, A., Coad, J., Plumridge, G. M., Gill, P. & Farndon, P. Family communication between children and their parents about inherited genetic conditions: a meta-synthesis of the research. *European Journal of Human Genetics*, 16, 1193–200 (2008).

Many parents reported that they felt they had a duty to promote openness and trust by communicating as a family which in turn enabled the family to be best placed to support each other and tackle any problems as they arose. This final point is highlighted by Anne, who thinks that the girls deserve to know:

“*I think the girls have a right to know, on the Yahoo forums² there’s a constant debate about whether they should know. I almost think it’s like their human right to know especially before they have to make major decisions about children and things.*”

Things to consider

Should I tell my child?

Reasons for not telling your child

- The consequences of Trisomy X can be mild or negligible.
- It can be stressful to be given diagnostic information that is hard even for adults to understand.
- Being told of the diagnosis could create anxiety and stigmatisation in the child.
- Could be used as an excuse for bad behaviour or not trying at school.

Reasons in favour of telling your child

- If your daughter is experiencing educational or emotional difficulties, it can be helpful to have an explanation.
- It can be uncomfortable to keep a ‘family secret’, and your daughter may be upset if she does eventually find out that information has been kept from her.
- Awareness of Trisomy X may empower your daughter, allow her to link up with others with the same condition and help her to make informed decisions in the future.

² Parental support group and forum (<http://groups.yahoo.com/group/raregeneticdisorders/>)

When and how to tell your child

If you do decide that telling your daughter is the right course of action for you, then the next big questions are ‘When should I tell?’ and ‘How should I go about it?’ The following sections provide insights from parents about both these questions.

Insights from parents on when to tell your daughter

Knowing when to tell your child can be difficult. When girls are still young, their understanding may be more limited, but they may be more accepting. Older girls may be better able to understand the complexities of the condition, but may be upset to discover that information had been kept from them until now. Some parents suggest waiting until your daughter starts to ask questions – which may, of course, never happen.

The experiences that follow are from parents who have told their daughter about having Trisomy X as well as from teenagers and women with Trisomy X. Peter and Anne told their daughter Abby after she reached puberty and Peter explains their reasoning for waiting until Abby was older and offers advice to other parents:

“*I think it's best to tell in the teenage years once they've got more genetics knowledge. It's meaningless for the child before then. Around then, our daughter asked if there was something wrong with her. For others I would say to tell when she's in her teens. Before then the distinction between peers isn't too great but when they get to teenage age it becomes more apparent.*”

Other parents also decided that telling their daughter later was the right decision for them. Pam explains about telling her daughter, Alexandra, when she was 14, but also highlights that other children may be less understanding in their teenage years:

“*I told Alexandra at 14 and her initial reaction was ok but then she told her friends and some of them decided to bully her about it but it all seems to have straightened itself out now.*”

Other parents, however, thought it was best to tell their daughters earlier on. As mentioned previously, Imogen decided to tell her daughter Natalie early on as Natalie was having difficulties at school. Imogen added:

“ *She has always known. We tried to tell her so hopefully she would try that bit harder! And why she needed special needs at school. She accepted that. Because she knew that she needed it, didn't she? Because children do accept things, they do.*”

Imogen raises an important point; that younger children are often more accepting. Additionally, telling your daughter when she is young allows you to build on her understanding year-on-year. In fact, some parents felt it was an advantage to start with explanations while the child was still very young, because it allowed understanding to develop gradually, with the child absorbing more as they grew older. Jason describes his decision to tell his daughter, Amelia, fairly early on:

“ *So we thought, number one hopefully she'll understand it, but I think, for us, it was a personal decision. And we did tell her, I can't remember what age we told her, but I think from whatever age she could understand and comprehend it. So she's always known. Yeah, so I'm guessing she was probably about six or seven years old when we told her. But she's always known.*”

I don't think there's much point in explaining it in too much depth from a genetic point of view because we don't understand it. We understand it broadly but nothing like you need to, it's a day to day thing that you need to cope with. Yeah, I think it was a personal decision for us to tell Amelia. And for us it was definitely the right thing to do. If you were asking me whether I would recommend anybody else to do it, I think it should be a personal decision.”

Although Jason decided to tell his daughter early on and chose not to go in-depth into any complicated explanations, he also raises another really important point – deciding when you think is best is a personal decision and, as a parent, you are ultimately best placed to decide when is the best time to tell your daughter.

Insights from parents on how to tell your daughter

Once you have made a decision about when to tell your daughter, the next decision to tackle is how to go about this. Even after committing yourself to telling your daughter about her trisomy, it can be very challenging to work out the best way to do so. Genetics is an exceptionally complex topic and many people find it confusing enough themselves, without having to work out how to explain it to a young child.

Additionally, there is the underlying concern that if you do tell your daughter that she has an extra chromosome; she may start to feel like a freak.

Some parents, who chose to tell their daughters at a fairly young age, approached the topic by simply telling them they were 'special' and not getting into the details about genes and chromosomes. Rachael describes how she decided to tell her daughter soon after she started school:

“ We told her that she was special, she might struggle a bit at school, but she’s special. She’s got this extra X chromosome and we worked it like that. We always called her ‘extra special’. We just kept it simple, to be honest.”

This approach was also favoured by Jason who decided not to introduce anything complicated about chromosomes but stick with how having Trisomy X affects his daughter Amelia day-to-day:

“ We sort of made light of it... We’ve always made a joke of it, ‘the XXX is coming out today, Amelia, is it?’ ‘it’s showing its head today.’ We’ve done the ‘special’ thing as well. I don’t think she needed to know any more than what we told her. Other than ‘you’ve got an extra female chromosome and it’s going to make you a bit more girly. You might be this, that or the other.’ We didn’t go into any more depth, because I don’t think she could have absorbed it to be honest.”

Sarah discusses plans to tell her daughter, Olivia, in a similar way by highlighting the everyday problems her daughter might face, particularly with regard to emotions. Sarah’s advice to other parents in similar situations is to get a feeling for when is the right time to tell:

“ That’s what we were thinking of doing; saying you’re a little bit more girly, a little bit exaggerated. So if you should be feeling a bit stressed, you might feel really stressed. Or if someone’s feeling a bit sensitive you might be extra sensitive. If I was advising anybody I would tell them to base it on how ... they’ve got a feeling really. It’s such a personal thing, I think. And it’s about the people you are around as well.”

Owen questions whether it is OK to just talk about the emotional side and not mention anything about educational difficulties and whether or not it would be fair to only give a one-sided explanation:

“ Say “you’ve got XXX,” but you don’t mention the educational difficulties, you just mention the emotional ones instead. So, “you have friendship difficulties,” or however you word it to a child, but you don’t say, “That’s why you are struggling with reading.” Or can you not give them half the story? Is that unfair?”

Where possible, parents felt it was important to stress positive as well as negative features of Trisomy X. One mother told her daughter that her ‘lovely long legs’ were part of Trisomy X that she could be proud of. A common point was that parents emphasised that Trisomy X was ‘not good, not bad, just different’.

Things to consider

Deciding when and how to tell your child

Your child’s actual age is less crucial than her level of understanding.

Advantages of telling your child while still young

- Avoids a sudden shock in teenage years.
- Limited understanding means the child can gradually absorb information as her understanding grows.
- Can use picture-book to introduce the idea of being ‘special’.

Advantages of telling your child when she is older

- She will have better ability to understand what she is being told.
- Allows parents to postpone decision to disclose diagnosis, especially if no problems encountered.
- “My Guide to Trisomy X” is designed to explain Trisomy X to older girls.

We have developed a booklet that can be used to help talk to your daughter about Trisomy X, incorporating some of the experiences of parents who helped with our research.

Using the “Things that make me special” picture book

The picture book “Things that make me special” is intended for younger children. We have done pilot studies to check the level of understanding required, and found that most children with a language level of at least 6 years can understand most of the content. Many girls with Trisomy X have immature language skills and social understanding, and so may need to be a bit older before they can comprehend

the main messages. Nevertheless, following what several parents have told us, it is possible to use the book with younger children, even if they understand only parts of it.

In the picture book we do not attempt any detailed account of chromosomes or genetics. Even the simplified account that we provide may be over the heads of many young children. However, our aim is that the picture book should nevertheless be useful in talking about differences between people, and the idea that Trisomy X is just one way in a girl can be different and 'special'. We also make it clear that, while some girls have associated difficulties, this varies a great deal from child to child.

As well as the main text, we have 'Talking Points' in coloured boxes, suggesting points that parents may wish to discuss with their daughter. This way, we hope it will be possible to personalise the way in which the picture book is used.

“My Guide to Trisomy X”

Our picture book can be useful for introducing the idea of Trisomy X to young girls, but it is limited in the amount of explanation it gives. There are some excellent resources available that give more information about the genetics and nature of Trisomy X. “My Guide to Trisomy X” is great for explaining Trisomy X to teenagers. It can be downloaded from <http://www.triple-x.dk/Erskineng.pdf>.

Deciding whether to tell brothers and sisters and how to go about this

Deciding whether or not to tell brothers and sisters can also be a difficult decision. Every family will have their own unique set of circumstances that will contribute to their decision to tell their other children and there may be many factors involved in making that decision.

In the first instance you have to decide whether to tell them before you have told your affected child and this decision will depend on your children's ages and the age gaps between your children. When families have an older sibling(s), particularly in the case of larger age gaps, telling the older sibling(s) first might be the most appropriate course of action. Melanie discusses her desire to balance wanting to wait and tell her youngest daughter, Rosie, that she has Trisomy X, with wanting to be able to answer her eldest daughter's questions about why her younger sister isn't meeting her milestones:

“ *The only person we're concerned about telling or not telling is her oldest sister (Kate) who's nearly 14. If she doesn't get told, you think is she going to say, 'Why didn't you tell me?' or if she does get told is she just going to worry what Rosie's going to turn out like.*

We do want to tell Kate because we think she should know because it's an important part of our family. But I think she would worry so much that we're always tempted to see how Rosie turns out and if she turns out to have ADHD at six or huge problems, then we can explain to our older daughter, 'She's got this XXX so please be a bit more understanding.' Whereas if she didn't have anything I think we'd tell them all at the same time.

She's a worrier, my older daughter, but if there were issues I think with Rosie, Kate is like, 'Why does she keep doing that, why is she like that? Why can't she keep up? Why can't she talk properly? Why can't she run properly?' Then we'd be, 'It's probably because she's got this.' And I think to have an explanation would make sense then to the older children.

So we're just waiting to see, because we've got a 12 year gap. So it's a big gap. If we don't tell Rosie until she's, say, six, Kate is going to be 18 and then she might be like 'Well why didn't you tell me before and I would have put more effort into helping my littler sister talk' or something."

Melanie's dilemma highlights that older children in the family may have already noticed that their younger sister is not able to keep up with her peers at school, or that she has additional behavioural or emotional problems and they may in fact ask questions about this. Asking about their younger sister may provide an excellent opportunity to talk to them about their sister's extra chromosome and why it might help explain her behaviour. In deciding whether to tell any older children about their sister having Trisomy X, you will also need to consider how your daughter might feel if she later realises that her siblings knew before she did. We've already noted how children with Trisomy X can be resentful if they feel their parents have kept information from them: this can be made even worse if they find out that brothers and sisters have been 'in on the secret'.

By not telling brothers and sisters, you risk leaving them to make incorrect assumptions about their sister's behaviour perhaps thinking that she is acting out for attention. Being open with other children about their sister's Trisomy X allows them the opportunity to provide help and support and will also help them to be more understanding and make allowances for poor behaviour. Jessica describes how useful telling her eldest daughter, Joanna, has been as she is now much more understanding of her younger sister, Lauren's behaviour:

66 *Joanna is ten years older than Lauren. I think in the beginning she was a little bit jealous of Lauren because more attention was focused on Lauren when she was younger – and I think she just thought she was being naughty – and because she's older she's a lot better with her – she helps her a lot more. And strangely enough, since Lauren has seen this other paediatrician who said 'I actually think she's got autism – she's on the autism spectrum' and having this XXX, it's like a professional has told her, 'your sister has got these problems' – and it's a bit of a wake-up call because she's got more patience with her. She helps her more.*

They have got much closer. She never used to like Lauren going in her room but now she'll have her in her room all the time. I don't know whether that's part of Joanna getting older and getting more sensible or she's just realising 'actually, my sister does need a bit more help and she doesn't always understand'. Because sometimes we will do things and Joanna will say 'she's just doing that mum'. And I say 'but she doesn't understand; you've got to explain things to her'."

Jessica's experience nicely identifies that siblings can often be closer to each other than they are to their parents and so making older siblings aware of their sister's chromosomal condition can allow them to retain that bond whilst also providing the younger daughter with someone else to talk about having Trisomy X. Libby also picks up on this and describes how useful it was to tell her son, Chase, about his sister, Aria, having Trisomy X:

“ We told our son when Aria was one. He started to ask why she was different, she seemed more immature than others her age, and he seemed relieved to know that there was a reason. He's very loyal to her, when we explained that he mustn't tell anyone else we know he won't as he's so loyal to her, he puts up with more from her now, before he would get frustrated but now he seems to make allowances.”

Libby's example does, however, bring out another aspect of disclosure: the need for discretion in telling others. Although some parents may feel that it does not matter who knows about their child's Trisomy X, many families would prefer that the information was not broadcast widely, because of the potential for stigmatisation of their child. But this means if another child in the family is told, they will need to be relied upon not to tell other people, and this need for secrecy can create a burden. Anne highlights the strong bond between her children when pondering her decision on whether to tell her son about his sister's Trisomy X:

“ We don't know what goes on in our daughter's head and if her brother knew then maybe he'd understand her better.”

If you have a daughter with Trisomy X who has younger brothers and sisters then you may choose to tell all the children about the trisomy at the same time. This can allow them to develop their knowledge of the condition together and will also avoid the issue of a sibling knowing about their sister's Trisomy X before your daughter does. On the other hand, if you prefer that information about your daughter's Trisomy X is kept private to the family, then it may not be a good idea to tell young children in the family, who may have difficulty keeping such information to themselves. This is obviously dependent on the age and level of understanding of the younger children. Jason describes his positive experience of telling his youngest daughter, Grace, that her older sister, Amelia, has Trisomy X:

“ Yeah, Amelia is two and a half years older. And I think from memory I think we told Grace about the same time. So Grace has always known. Grace has always been a little bit brighter than Amelia so she's always understood it. And it's definitely been a help to Grace because she does get a hard time from Amelia. She always has done. So she's got something to blame

it for, because otherwise I think she would have taken it personally, more personally. Well we all would have, we all get a hard time from Amelia when the XXX is coming out. And if you didn't have something to blame it on, it would be more difficult to ... you would definitely take it more personally. But we did tell Grace at the same time that we told Amelia which would have been when Amelia was about seven."

Jason's experience highlights another reason to let siblings know; telling them provides them with an explanation for their sister's behaviour. Siblings can often be mean to each other, though if a sibling is on the receiving end of particularly bad behaviour or emotional outbursts from their sister that may be due to her Trisomy X, then by telling her siblings, they at least know that their sister may be acting out because of her Trisomy X and can be more understanding. By knowing that there might be a reason for their sister's behaviour, they may be able to deal with her behaviour better.

You may decide that telling siblings may not be necessary or appropriate if your daughter with Trisomy X is not showing any noticeable problems or difficulties. Sarah discusses her decision and offers a perspective from a family who have another child with a developmental disorder and her reasons for not telling her other children about Olivia having Trisomy X:

“*My scenario is slightly different because mine are the three boys; they are younger than Olivia. So I've got their levels of understanding ... so I think when we tell her we probably won't tell the younger ones and then if the teenage years perhaps are troublesome, it might be then that we choose to tell them. I've not got any preconceived ideas on that, actually. I think it's a wait and see for us. So with my son having ADHD, he's got enough to cope with without worrying about his elder sister. But if she's going to have difficulties I would like for her younger brothers to look after her sort of thing when they are that bit older. So we'll see what they turn out like."*

Sarah makes an important point that if it may not be useful for siblings to know if your daughter with Trisomy X is not experiencing any problems at home or at school. In this case, waiting to see might be beneficial, though, if you later tell your daughter, be prepared for other siblings to wonder why you didn't tell them earlier.

Things to consider

Telling brothers and sisters

Reasons to tell brothers and sisters

- Other children might be able to provide support to your daughter with Trisomy X and help her if she struggles with anything.
- Siblings might be more understanding and accepting of any poor behaviour.
- Siblings might provide an alternate place for your daughter to turn to for support.

Potential problems in telling brothers and sisters

- Children may find it a burden to keep a secret.
- Children with Trisomy X may be upset if they realise others in the family have known information that was kept from them.

Factors affecting when to tell brothers and sisters

- The age between your children might determine who gets told first.
- Telling siblings at the same time can allow for their understanding to grow together.

Deciding how to tell brothers and sisters

- Think about whether you want to wait and see if they ask questions or find an opportunity to bring it up.

Deciding whether to tell your daughter's school

Deciding whether or not to tell your daughter's school can be important, particularly if she is experiencing any difficulties and would benefit from support at school.

For most parents in our focus groups, a decision to tell the school was prompted by awareness that their daughter would benefit from extra support. Parents tended to avoid telling the school about Trisomy X if they felt their daughter was coping OK, and so nothing would be gained by disclosing.

Rachael describes why she hasn't informed her daughter, Savannah's, school about her having Trisomy X yet:

“ *And I haven't told the schools, only because I don't think she needs extra help, which I do sit with her doing extra help, but up until now at school she's coped okay – she puts a lot of pressure on herself and really wants to do well but sometimes she just can't do it and then she has this kind of block that comes on 'I can't do it' ... especially with maths – and that is supposed to be the issue. And she really struggles. And then you just try and do it. So I got her to doing extra work ... there's enough books out now or there's the internet or maths programmes – doing fun stuff – and she's actually done ... she's still at the lower end of the class but she's there and she's coping for now.”*

Where children are experiencing educational or emotional problems, there is a stronger motivation to tell the school. This is balanced, though, by a concern about possible stigmatisation and lowered academic expectations. In practice, parents' experiences were hugely variable, and the response from school staff ranged from very positive to totally negative. Imogen relates a more negative experience:

“ *And so in a way I think if I hadn't told the school, would she have got more time spent on her, rather than, “Oh well she's never going to learn,” so as long as she reads the Sun newspaper, let's not worry? That was a decision I went into thinking that they were going to help me. We thought we should have waited to see really because we didn't want people looking at her and going. “Oh, poor little thing,” and thinking the worst when we hadn't given her a chance to see if she was going to be affected by it or not.”*

In stark contrast, Karen explains how her daughter's nursery was very accommodating and willing to work with her daughter's individual needs:

“ When I told the nursery – they’re brilliant at our school anyway, she said ‘oh that will be really interesting, we’ve had a child with Fragile-X before but I’ve never seen XXX so I will really keep you posted on how she’s doing’. She said, ‘if you wanted, start her at three days but then if she can handle five days then we’ll bring it on ... or, put down five days because you’ll probably have signed in for it and if she doesn’t feel like coming in, ring up’. They were really flexible which was good. But she went straight on in there. She was not fazed by going into the room with all the other children or anything.”

Considering that different schools may react differently is important. Use your judgement to decide how you think the school would react to the news. For example, if the school have any other children with disabilities already in attendance this might be positive and you can be sure that the school are able to cope with children who are different, and that other children will already have an awareness of individual differences. Karen goes on to explain that the school her eldest son, Michael, attends is brilliant with children who have disabilities and that she feels confident that when her youngest daughter starts at the school, she will tell them about her having Trisomy X:

“ We’re quite lucky because with the school that Michael goes to – they’ve got a lot of special needs – so they’ve got children with cerebral palsy, they’ve got autism – and so when Caroline will hopefully be going to school, they’re all aware of that big picture of differences.”

If your child is in need of support at school it is worth remembering that help might not always be easy to come by and that you might have to fight for it. Jane explains how, when it became obvious her daughter was falling behind her peers, she told the school about Abby having Trisomy X and expected more support than was offered:

“ I didn’t mention it at school until it became apparent that she wasn’t achieving as well as others, but we weren’t offered any help. She was a low achiever but she didn’t cause trouble so she didn’t get help, I had to fight for it and am seen as somewhat of a ‘problem parent’. She has issues with social groups and friendships and they gave her some help there, but at GCSE the gap was getting wider and we got her statemented, but now they won’t take her on for A Levels so she’ll have to leave all her friends to go to Sixth Form College.”

Jane's experience mirrors that of many parents whose children have difficulties at school; although the school is aware there may only be so much that they can do. One point stressed by parents was that it was extremely unlikely that school staff would know anything about Trisomy X, even if the school was familiar with other disabilities. If you do plan to tell the school, go prepared with material that explains the diagnosis, such as the Unique document "Triple X."³ Jessica explains the reaction of Lauren's teacher to being told she has Trisomy X:

“ I think when we go to school we told them about Lauren's XXX ... of course the teacher says 'never heard of it' so he prints stuff off and there's pages and pages and pages and they sort of read the first page and it gets ... 'whoosh'. We've been struggling with the school for the last five years to get her help.”

Although the medical-sounding label Trisomy X may get your child's problems taken more seriously than would otherwise be the case, the label alone does not really provide any guidance to the school as to how to help. Even if teachers read up about the condition, they will come away uncertain, because the educational profile varies so much from child to child. The diagnosis may help you get an assessment for your child, and will also indicate which areas of functioning are likely to pose problems, but on its own it is not very informative. Do not expect, therefore that the diagnosis alone will lead a school to recommend a particular type of support. It will be necessary to get an assessment by a professional such as an educational psychologist or speech and language therapist. This will allow school staff to characterise the child's profile in terms of difficulties they are familiar with. Trisomy X is a risk factor for developmental difficulties, especially those affecting language and social interaction, but your child's personal profile of strengths and difficulties needs to be identified.

Jessica explains how her daughter receiving a diagnosis of autism spectrum disorder will be more likely to get her support at school:

“ She needs extra help because we went to a paediatrician who has also said now that she's on the autistic spectrum. And so we're hoping now that this report will go towards getting a Statement [of Special Educational Needs]. She was referred to a communication disability team – a lady went into the school and spent some time with her and noted everything that the teachers have to do with her.”

However, it's not all bad and Jessica further explains that through working with the school for help for her daughter there are now measures in place such as the school having a person in place that Lauren can go to if she needs to for any reason:

³ <http://www.rarechromo.co.uk/html/DisorderGuides.asp>
See also: Useful resources at end of booklet.

“*I’ve stressed ‘it may be difficult but please just try and walk away and find an adult’. So they’ve assured me now that there is a person that she can go to if these things do happen.”*

Things to consider

Telling the school

Deciding whether to tell the school

- If your daughter is not experiencing any difficulties, then telling the school about Trisomy X may do more harm than good, by creating low expectations.
- If your daughter needs help and support at school, then a Trisomy X diagnosis may be helpful in getting her needs taken more seriously. It’s important to remember, however, that even after telling the school you may still need to fight for your daughter to get support.
- Schools that have a good track record with children with disabilities might be more understanding and more accommodating of any specific needs.

Deciding how to tell the school

- Print off some information about Trisomy X and be prepared to explain a bit about it to the school.
- Discuss the specific difficulties that your daughter is experiencing, and ask for a professional assessment of her educational needs.

Deciding whether to tell other family members and friends

Telling other members of your family or confiding in friends can provide a vital support network for your family, though there may be other concerns such as not wanting any pity or for your daughter to be pre-judged.

One of the main concerns many parents raised was that their daughter might be pre-judged by family or friends before they had got to know her or waited to see whether she had any difficulties. Owen explains that, so far, he hasn't told any family members or friends as he doesn't want them to think his daughter, Aria, won't be able to achieve anything because she has Trisomy X:

“ We've never mentioned it. We're just going to wait and see with family, aren't we? Because otherwise I think people will judge her straight away and knowing what we were told, if we just said, "Oh she might be like this," they would expect her not to achieve much. And that's not an option really. We didn't want the family to view our daughter as a disabled baby; we want them to get to know her first.”

Julia also explains that she hasn't told many friends as they might think that her daughter having Trisomy X was an explanation for any of her bad behaviour and that she doesn't want her to be labelled in any way:

“ I haven't even told any of my friends. There's only one friend who's got a son in another school, and she knows. So I could phone her but I haven't told anyone else ... it's that thing when you tell somebody and you hope that they are going to be nice and kind but if she did misbehave or has a crying moment, 'oh look at her, it's because of the XXX'. It's become a label in itself.”

This point of view was also shared by other parents including Jessica, who agreed with not wanting her daughter to have a label that singled her behaviour out as different:

“ I tended not to tell a lot of my friends because I didn't want 'oh let her get away with bad behaviour because she's XXX' and I thought, no, she might be XXX and she's got a label, but I wanted her to be treated normally.”

Other people's ignorance of the condition can work in your favour. Rachael explains that when other people haven't heard of Trisomy X, they will only know what you tell them:

“ But I think everyone's ignorance of the condition actually has benefited because they've got no preconceived ideas. There are a lot of people who go, 'Really, is there something wrong with her?' Which is a kind of nice thing to say; 'Really, I would never have thought it.' ”

One mother, Imogen, described not wanting any pity; after trying for a baby for many years, she didn't want her happiness tarnished with people feeling sorry for her:

“ Well, mine was purely selfish because I'd been trying for so long for a baby, I didn't want, 'Oh poor Imogen, she's tried so long and there's something wrong with her baby;' so I didn't tell anyone straight away. We didn't tell my mother-in-law either. If you think people are going to understand or there's a benefit for you to tell someone, like the school, then yes. But we haven't really told many people.”

Some parents explained that they were very selective in deciding who to disclose to and that the people they've told have been told because knowing is of some benefit. David and Melanie took this approach. David explains his position on telling other family members and friends:

“ For us we've been selective in who we've told. We've not told Melanie's parents, her parents don't know. Some of our friends do know and certain don't. And I think it's been based on the personalities of those people really. Some that we knew would understand. One of my wife's friends is a nurse, so she knows. And she understands it from a medical point of view, and is quite interested in it as well from a medical point of view. My wife's closer friend, she doesn't know. But no reason for not telling her, other than it wouldn't bring any benefits to the situation. So we've been selective based on the people that we know and their personalities really.”

David's decision to base who they tell on any perceived benefits seemed fairly common and other people similarly only disclosed to others where they received a direct benefit such as support. However, some parents thought that being more open was the way forward. Karen described her view on telling others:

“*We're the complete opposite because I can't not tell anyone. I don't know what it is but I'm not being apologetic or anything because she doesn't do anything wrong, but I don't know ... my family are pretty useless – I have to talk to somebody so my closer friends all do know about it. You almost have to educate other people in order to de-stigmatise it.”*

Karen's experience not only provides her with necessary support from friends, but by being open she is raising awareness of Trisomy X and not allowing anyone else to pre-judge her daughter.

Things to consider

Telling family and friends

Deciding whether to tell family and friends

- Family and friends can be a great source of support and choosing to tell them can provide you with someone to help you through any difficulties.
- You may want to consider being wary of stigmatisation, although most people are ignorant of the condition, and so will only know what you tell them about Trisomy X.

Deciding when to tell the family and friends

- Are friends and family asking questions? This may happen if your daughter is clearly having difficulties or not meeting milestones and this can provide an opportunity to let them know about the Trisomy X.

Deciding how to tell the family and friends

- Think about telling others about the difficulties your daughter faces or going into more details about the genetics of the condition.
- If family and friends ask about your daughter, this could be an opening to tell them about her Trisomy X.

Special thanks to Prisca Middlemiss, Sarah Wynn and the staff at Unique for their support in organising the Study Days, to Karen Melham for assistance with focus groups and to all the families who gave up their time to talk about their experiences, without whom this would not have been possible. We would also like to thank Jessica Myring, Debbie Shears and Diana Wellesley for their support and advice in putting together this guide.

This guide was produced by Nikki Gratton, Dorothy Bishop and Gaia Scerif of the University of Oxford.

This project was funded by the Nuffield Foundation, although the views expressed are those of the authors and not necessarily those of the Foundation.
www.nuffieldfoundation.org



Oxford Study of Children's Communication Impairments (OSCCI)
University of Oxford
Department of Experimental Psychology
South Parks Road
Oxford, OX1 3UD