Talking with your child about his diagnosis of XXYY syndrome

Many parents wonder how and when they should tell their son about his diagnosis of XXYY syndrome. This guide offers some suggestions for talking with your son about XXYY. As part of a research study, we asked adults and parents of children with X or Y chromosome variations about their experiences discussing the diagnosis. We also asked what advice they would give other parents who are planning to talk about the diagnosis with their son for the first time. This guide was developed from their responses, as well as from recommendations by healthcare professionals.

Why is talking about the diagnosis important?

There are many reasons why talking about the diagnosis is important for your son and your family:

- Children with XXYY often experience speech, learning or social challenges starting at a young age. They may feel different from their peers. Having information about the diagnosis can help your child to understand and accept his differences.
- Your child should hear the diagnosis from you, or from a support professional (like a doctor, therapist or genetic counselor) with you there. Your son can then address questions and concerns with you when he has them. Otherwise, your child may learn of his diagnosis incidentally. For example, he may overhear it in a conversation, or be told by a healthcare professional or teacher who assumes your child already knows. This may cause feelings of betrayal and misunderstanding.
- Children are intuitive and often aware when something is being kept secret. They may imagine something much worse than their diagnosis, like that they or a family member has a serious illness.
- Secrecy or reluctance to talk about the diagnosis may make your child feel that XXYY is shameful or embarrassing. If your child feels that you are uncomfortable talking about XXYY, he may avoid asking you questions. Instead, he may seek information from sources that could be unreliable or unsafe.

Common Parent Concerns

- It is normal for parents to have concerns about telling their child about his diagnosis. You may be worried that:
- Your child will think that he is different or that there is something wrong with him, or that it will lower his self-esteem.
- You do not know enough about XXYY to explain it correctly, or to answer questions your child might ask.
- Your child will use his diagnosis as an excuse not to try when he faces a challenge.
- It will make your child feel upset, scared, or angry.
- Your child will not understand the information.
- Your own emotions will interfere with talking to your son about his diagnosis.
- Your child will not understand when it is and isn't OK to tell other people about his diagnosis.
- You will have difficulty talking about sensitive topics such as learning disabilities, pubertal differences, or fertility.

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How will my child respond?

Your child's maturity level, personality, and symptoms will affect the way he responds to the diagnosis. Keep in mind that your son's feelings towards the diagnosis may change over time. Your son may:

- Feel relieved
- Want to know who else knows, or when the diagnosis was made
- Need time to think about it
- Have a neutral reaction, seem disinterested, or act like it is "no big deal"
- Feel sad or angry
- Feel confused
- Feel special or think it is "cool"
- Wonder why it has happened to him
- Watch to see how you feel about it
- Ask questions about what the diagnosis means for him or how it will affect his future
- Show his emotional response through his behavior or actions, such as acting out or a change in appetite.
- Have a delayed reaction. For example, your child might seem disinterested at the time of disclosure, but distressed the next day. Or, he may ask more questions later.
- Have varied reactions to different aspects of the diagnosis. For example, your son might feel anxious about hormone treatment, but relieved to know why he is struggling socially or in school.

When is the right time?

Many parents wonder about the "right time" to bring up the diagnosis. Here are some considerations for when to begin the conversation about XXYY with your son. Remember, there is no specific "right" age, and every child is different.

- Keep in mind that there is not likely to be a "perfect moment" to bring up the diagnosis for the first time.
- For children diagnosed prenatally or in infancy, it can be appropriate to first mention the diagnosis around age 4 or 5. For boys diagnosed in childhood or adolescence, discussion may start at the time of diagnosis.
- Even if your son is young, it may be helpful to talk about XXYY a little at a time as relevant to his age and symptoms.
- Your son should be aware of his diagnosis before he begins puberty and will need to start testosterone therapy.
- Rather than create an event, like going out to dinner or a special outing, try to find a casual time for the disclosure conversation.

Other opportunities to discuss XXYY include:

- If he experiences learning or social challenges
- When he brings it up
- Preparing for appointments (i.e. doctor, therapy, tutor)
- If he needs medications or treatments
- XXYY Conferences or family meetings
- If he is learning about related concepts in school (i.e. chromosomes, human body, cells)
- Books, movies, or TV shows with related topics

Preparing for the conversation

- Become informed about XXYY before you talk with your child. Seek answers to your own questions from doctors, other parents, and reliable websites and books. This may help you to feel more confident when talking with your son.
- Children often pick up on their parent's emotions. Take the time you need to work through your own feelings about the diagnosis and about talking to your child. You may want to seek support from family members, friends, a professional therapist, support groups, or community groups of other parents of boys with XXYY.
- Consider talking to your child's doctor or therapist about your plans to disclose the diagnosis. He or she may have advice on age-appropriate ways to talk with your son. A healthcare professional can also help you to disclose the diagnosis to your child, or to talk to your child about specific aspects of his condition.
- If your child has more than one parent or caregiver, discuss together ahead of time how and when you will talk to your son about his diagnosis.

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What Should I Say?

Your son's unique needs and your family communication style will influence how you discuss your son's diagnosis with him. Some points to consider:

- Keep it simple - most children need minimal information at the start
- Be honest and direct. Do not give false or misleading information that may cause him to be confused.
- Keep a light, calm, and positive attitude.
- Encourage your child to ask questions and to share what he is feeling or thinking, acknowledging he may need time.
- Express support and reassurance. Offer to help your child get any help he may need for the challenges he may experience.
- Relate your son's condition to others' differences or challenges, with examples relevant to your child and your family. For example, testosterone therapy may be compared to a friend who requires insulin for diabetes, or thyroid hormone treatment.
- Consider using pictures, blocks, or other visual aids when talking about complex topics like chromosomes or testosterone.
- Identify your son's strengths and positive qualities. Also try to identify good things about the diagnosis (such as it has helped your son be tall, artisitic, or has made him more sensitive to others).
- Let your son know there are a lot of other boys and men with XXYY.
- It is ok if your child has questions that you are unable to answer. Become comfortable with telling your child, "I don't know, but we can find out together."
- Tell your son that it is always ok to ask about anything he may read, hear, or wonder about. Let him know that many things he reads or hears about XXYY may be wrong.
- Help your son identify appropriate places to seek information about his diagnosis, such as his doctor, safe websites or books. Encourage him bring any questionable information to your attention.

Developmental Considerations

Remember that you do not need to talk about everything at once. The topics you discuss and the words that you use will depend on your child's age and experiences. These are some age-appropriate suggestions:

Preschool/Kindergarten:

- Explain that your son's body and brain are made a little differently than other children, and that this means that he might need more help (i.e. speech therapy) or that some things may be harder for him than other children.

School age:

- Begin talking about chromosomes and genetics, providing more details as your son learns about these topics in school. Reiterate that some information he learns at school may be wrong, and to talk to you if he has questions.
- Reassure your child that XXYY is a condition they were born with. It is not a disease, and that it is not contagious or fatal.

Adolescence / Adulthood:

- Talk with your son about his feelings about sharing his diagnosis with friends and romantic partners.
- Introduce the fertility problems associated with XXYY (see tips on next page).
- Discuss ways that your son can become proactive and involved in his health care. Encourage him to ask his doctors questions.

XXYY versus "Klinefelter syndrome"

Klinefelter syndrome is the medical term used to describe a more common condition of 47,XXY in males. XXYY is sometimes called a "variant" of Klinefelter syndrome. There are many medical and developmental issues that are common in both XXY and XXYY. However, it is important for people to understand that XXYY is usually more complicated and more likely to be associated with disability than XXY, and so it is important that people understand how XXYY is different than XXY (Klinefelter syndrome).

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Talking about privacy

- It is important to discuss with your child who is appropriate to tell about the diagnosis, and who may not be appropriate.
- Tell your child that sharing the diagnosis is a personal decision to be made by the child and his family. Most people will not understand what XXYY means.

Talking about genetics and chromosomes

- Chromosomes and genes may be explained as "instructions(or messages) for the body," or as "pages in an instruction book." XXYY is caused by "extra instructions" or "extra pages."
- It is important to emphasize that XXYY is something that "just happens" and that he was born with this; no one caused your son to have it. Avoid using the words "mistake," "accident," or "mutation" to describe this condition.
- Be careful when using the terms “sex chromosome” or “sex chromosome abnormality.” Your son may confuse these terms with having a “sex abnormality.” Older boys or adolescents may need to be reassured that their diagnosis does not imply inability to have sex.

Talking about infertility

- From a young age, emphasize the different ways families are made, such as though adoption or using sperm donation.
- Address specific questions and concerns regarding infertility as your son matures. Waiting until adolescence to discuss fertility issues is appropriate.
- Teens should be informed that protection is still necessary to prevent sexually transmitted diseases.

Sources for this guide include:

- A Dennis, S Howell, L Cordeiro, N Tartaglia, ’How Should I Tell My Child?’ Disclosing the Diagnosis of Sex Chromosome Aneuploidies. Journal of Genetic Counseling, 2014 Sep 3
- Tartaglia, N., Ayari, N., Howell, S., D’Epagnier, C., Zeitler, P. (2011). 48,XXYY, 48,XXXY and 49,XXXXY syndromes: Not just variants of Klinefelter syndrome. Acta Paediatrica, 100, 851-860.
- Tartaglia, N., Howell, S., & Bornstein, A. How to talk to your child about his or her diagnosis. Presentation at KS&A Families Conference (2008)
- ACT Government Parentlink. Donor Conception (Telling your child). parentlink.act.gov.au

Remember:

- Talking about the diagnosis is not a one-time event. It is a process that will happen gradually over time.
- Every child with XXYY is different with regard to his needs, comprehension, communication and coping skills. You know your child best.
- Consider seeking support for talking with your child about his diagnosis from your child's physician, therapist, or other healthcare professionals.

Resources:

AXYS (www.genetic.org)

The XXYY Project
(www.xxyysyndrome.org)

American Association for Klinefelter Syndrome Information and Support (AAKSIS)

www.aaksis.org

Book: Living with Klinefelter Syndrome (47, XXY), Trisomy X (47,XXX) and XYY

by Virginia Isaacs Cover (2012)