

FRAGILE X INFO SERIES: SIBLINGS



Overview

Fragile X is a group of conditions associated with changes in the Fragile X gene – called FMR1 and located on the X chromosome. The FMR1 gene can undergo changes, when inherited, which affects a pattern of DNA called CGG repeats. Typically, the FMR1 gene has up to 54 CGG repeats, though the range between 45 – 54 repeats, is called the intermediate or gray zone. A premutation carrier has 55–200 CGG repeats, and someone with a full mutation has more than 200 CGG repeats.

When a premutation or full mutation is present, it can result in a Fragile X-associated Disorder (FXD). These include:

- Fragile X syndrome (FXS): A condition affecting intellectual, behavioral, and social development. It occurs in both males and females who have a *full mutation* of the FMR1 gene.
- Fragile X-associated tremor/ataxia syndrome (FXTAS): An adult onset (over 50 years of age) neurological condition, seen in males and females, but more common and more severe in some male *premutation carriers*. It can cause tremors, memory, and balance issues.
- Fragile X-associated primary ovarian insufficiency (FXPOI): A condition affecting ovarian function that can lead to infertility and early menopause in some female *premutation carriers*.
- Other issues may be present in *premutation carriers*, and this is an ongoing area of study for researchers.

The Sibling Relationship

The sibling relationship can be one of the closest and most enduring relationships in a person's life, leading to countless rewards, support, and growth. Developmental disabilities in one child, however, can present unique challenges for unaffected siblings. Just as with other disabilities, siblings of children with FXS may feel torn between two sets of feelings. They may feel embarrassment, guilt, isolation, increased responsibility, and pressure to succeed. On the other hand, their experience may also help them develop uncommon maturity, insight, tolerance, pride, and loyalty. They may even feel drawn to a vocation related to disabilities as a direct result of the relationship.

Growing up with siblings who appear different can be difficult. Explaining unusual behaviors to their peers, such as why a brother suddenly begins to hum or flap his arms, is not easy. Conflicts can also arise about fairness and why parents tolerate certain behaviors in an affected sibling, but not in the others.

Having a sibling with FXS can affect most everything the family does. Trips and family outings may get cancelled due to the behavior of their sibling. Many unaffected siblings grow up witnessing trips to hospitals and clinics and observing their parents' frustration as they seek answers about their sibling. Counseling for siblings not affected by FXS may be as important as for those who are affected.

Developmental Changes

Clinical studies suggest siblings' feelings follow a developmental pattern:

- Ages 1-5: Competition for parent time, jealousy, imitation, attention-seeking behavior.
- Ages 5-10: Resentment, friendship, caregiving, sense of loss.
- Ages 10-15: Embarrassment, anger, excuse-making, sorrow, and defensiveness.
- Ages 15-20: Caregiving, self-motivation, life choices affected by the relationship, proving competence.
- Ages 20 and older: Self-denial, estate planning, financial obligations, responsibility to maintain family ties.

Factors in Premutation Carriers

Premutation carriers are perhaps the most acutely impacted by their relationship with siblings who have FXS. Not only must they meet the challenges posed by the sibling relationship, but they also face the possibility that their sibling symbolizes their future as a parent of a child with FXS. They may consider reproductive counseling about the transmission of the Fragile X mutation.

Siblings Without FXS

Siblings not affected by FXS and who do not carry the gene mutation may experience a type of “survivor’s guilt.” They may ask, “Why did I inherit a normal X chromosome while others in my family did not?” They may be more likely to feel an obligation to help care for the sibling and pressure to succeed. Others worry about the responsibility they may face when their parents can no longer fill the caregiving role for their sibling.

Supporting Siblings

Peer groups are often helpful for siblings of children with FXS. It is a chance to meet others in similar situations and share their experiences in a safe environment. See the resources below. Some siblings may benefit from regular therapy with a trained professional.

Aggression

Approximately one-third of individuals with FXS exhibit aggressive behavior. Siblings are sometimes the target, and they may experience anger toward their affected sibling. While they may or may not understand the cause of their sibling’s behavior, they need to feel safe themselves. This can be difficult and confusing. Parents can recognize the potential for conflicting emotions and help siblings identify and verbalize their feelings. They need to know it is not acceptable for their affected sibling to act aggressively toward them.

Empowerment Ideas

For Parents

- Provide siblings with ways to manage the stress of having a sibling with FXS. Possibilities include—
 - A private area in the home.
 - Headphones for music to reduce environmental noise.
 - Regular, planned activities that are not related to FXS or their sibling, such as sports, dance, or horseback riding. Even a day trip with just the two of you can be a way to stay connected.

For Siblings:

- Consider activities to educate friends and teachers at school, e.g. preparing a presentation on FXS or on certain characteristics of those affected by it.

About the NFXF

The National Fragile X Foundation (NFXF) was founded in 1984 to support individuals with Fragile X syndrome (FXS), their families, and the professionals who work with them. Today, it is a comprehensive resource not only for FXS, but also for the conditions of Fragile X-associated tremor/ataxia syndrome (FXTAS), Fragile X-associated primary ovarian insufficiency (FXPOI), and other premutation carrier conditions and disorders. The NFXF is dedicated to serving the entire Fragile X community to live their best lives by providing the knowledge, resources, and tools, until, and even after more effective treatments and a cure are achieved. Learn more at <https://fragilex.org/welcome>

If you have questions please reach out to us at treatment@fragilex.org or call (800) 688-8765.

Resources:

Sophia James: Life as a Sibling of Someone with Fragile X Syndrome

<https://fragilex.org/international-fragile-x-conference/sophia-james-life-as-a-sibling-of-someone-with-fragile-x-syndrome/>

Sibling & Self-Advocate Network

<https://fragilex.org/living-with-fragile-x/community-support/sibling-self-advocate-network/>

Talking with Your Children about Fragile X: Why, How, and When?

<https://fragilex.org/fxs/talking-with-your-children-about-fragile-x-why-how-and-when/>

