

**What is FOP?  
Questions and Answers  
for the Children**

This book was written by Sarah Steele and her mother Marilyn Hair. The "I" voice in this book is the voice of Sarah Steele.

**Recommended for ages 8-12. For younger children with adult supervision.**

If you count how many times one of these words is in this book...**probably, usually, sometimes, some, seems to, often, might, could**...it would "probably" be 100 times. That's because every person who has FOP is different. Flare-ups aren't all the same. Some people grow more FOP bones than others. But even though we're different in some ways, we have a lot in common.

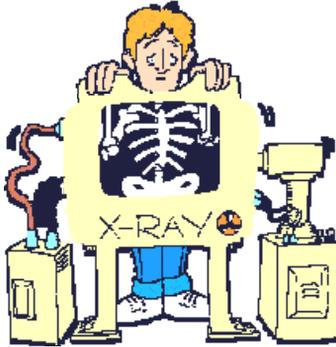
The authors hope that this book answers some of your questions and gives you ideas about where to find more help as you adjust to living with the mysterious condition we call FOP.

## What is FOP?

FOP stands for *fibrodysplasia ossificans progressiva*, which is a very complicated name to remember.

FOP is a disease that causes extra bone to grow in a person's body.

FOP bones are regular bones. They're just in the wrong places. They grow in your muscles, like in your back, shoulders, arms, and legs. They also grow in "connective tissue," which connects your muscles to your bones. The FOP bones make it hard to move. You could say that your body is growing another skeleton, an FOP skeleton, along with the regular skeleton you were born with.



## How did I get FOP?

You were born with it. If your mother or father has FOP, you got it from them. But if they don't have it, then FOP was an accident of nature. Scientists call it a **mutation**. But I'm getting ahead of myself.



FOP is a genetic condition.

(You learn a lot of new words when you learn about FOP).

Every living thing has a set of instructions in its body called **DNA**. DNA works like a computer program. A computer program tells a computer what to do. Well, DNA has instructions which tell your body what to do.

These instructions are called **genes**.

There is a gene which tells your eyes to be **BROWN** -- or are they **BLUE**?

Other genes decide what color your hair is, and how tall you'll grow. When you have FOP, one gene in your DNA is telling your body to make bones in your muscles.

So, how did you get FOP?

Your mother's egg and your father's sperm came together to make you. Your mother's DNA was in the egg and your father's DNA was in the sperm.

The two sets of DNA joined together to make a new person-- you!

DNA is the reason that some things about you are like your mother and some things are like your father. If your parents don't have FOP, one tiny part of the DNA they put together changed. Your FOP was a mutation or change in the DNA you got from your parents. It just happened. It's nobody's fault.

If your FOP is because of a mutation, it's almost sure that your brothers or sisters won't have it.

Also, FOP is not contagious. Nobody can catch it from you.

## What does it do to your body?

FOP kind of gives you a second skeleton.

When you're born, your big toes show you have FOP. Your big toes may be short and curved toward the other four toes.

Your parents and doctors noticed the shape of your toes when you were born, but they probably didn't know it meant you had FOP. Some people with FOP have thumbs that are shorter than most people's.

You don't have FOP bones when you are born. They grow as you grow. Usually FOP bones start growing in your neck, back (spine), and shoulders. Later they can grow in your hips, elbows, knees, and jaw.

The muscles in your heart, stomach, diaphragm (a muscle in your chest that you use to breathe), intestines, face, and eyes don't grow FOP bones.

One bad thing that FOP bones do is get in the way of joints. Joints are the places where two bones come together and move, like at our shoulders, elbows, hips, and knees.

If FOP bones grow in your shoulder, you can't raise your arm because FOP bones are in the way. If they grow in your knee, you can't bend your knee and that might make you walk with a limp.

People who have FOP get FOP bones in different places in their body when they are different ages.

My elbows were one of the first joints to grow FOP bones. When I was one year old, my elbows got stiff until I couldn't open and close my arms. But I know a man who has FOP who has been grown up for a long time. He can move his elbows almost as well as somebody who doesn't have FOP.

Different people who have FOP also have joints that are stuck in different positions.

For example, my elbows are closed so I hold my hands in front of my chest. Another girl with FOP has one elbow that's stuck wide open. She holds her arm straight down by her side.

Some people have more FOP bone in their body and more joints that are stuck than other people with FOP. Nobody knows why it happens this way.

FOP bones can also grow in muscles that aren't close to joints. Sometimes they cause lumps that stick out a little under your skin. It might happen on your back or on your head.

The lumps are normal bones that have grown in places they don't belong. The lumps won't go away, but they can change a little in size and shape so sometimes they seem to get smaller.

## Does FOP hurt?

It doesn't hurt all the time. It does hurt during a **flare-up**. That's when FOP bones start to grow. A flare-up happens in one part of your body; let's say it's your arm. First you get a **lesion**. A lesion is a swelling under your skin. It can be as small as a coin or as big as your whole upper arm. The lesion may feel warm. It might hurt to touch it and when you move your arm. I find it more comfortable if I don't move too much when I have a flare-up. I spend more time in my beanbag chair and in bed. The pain from a flare-up can last for a couple of weeks or more. Then the lesion gets smaller and it usually stops hurting.

It take anywhere from several weeks to several months for bone to form and after that you might feel a hard lump. Or maybe you won't be able to move your joint as wide as you could before. There isn't any medicine yet to stop FOP flare-ups, but there is medicine to keep it from hurting so much. Talk to your parents and your doctor. They can find medicine to make you more comfortable.

Sometimes people wonder if the weather affects FOP. Does it hurt more on rainy days, or when it's cold outside? Some diseases do hurt more in certain kinds of temperatures, but FOP isn't like that. People who have FOP might prefer warm, sunny weather just like anybody else, but FOP doesn't get more uncomfortable because it's rainy or cold.

## Will it get worse?

Yes, unfortunately, FOP does get worse.

The "P" in FOP stands for "Progressiva." That means that FOP progresses, or gets worse, as you grow up. The FOP bone that grows in your body doesn't go away, and more FOP bones keep growing as you get older, so it gets harder and harder to move.

**There are some things you can do to protect yourself.**

FOP often gets worse at a place on the body where there's trauma. Trauma means getting hurt, and it can happen from falling down, getting a bruise, surgery, or shots.

FOP is a funny disease. Sometimes FOP bones grow when you have the smallest bit of trauma to your body. But sometimes you can get badly hurt and no new FOP bone will grow. There are even times when FOP bones grow and you didn't have any trauma at all.

**Try to remember that getting a flare-up is not your fault.**

You can't make FOP start and stop. And since there's really nothing you can do to stop FOP bone from growing, try not to be so afraid of getting hurt that you don't try anything new or fun. FOP is only one thing about you. You don't have to let it run your whole life!

## What things could make my FOP worse?

Don't be afraid to do every little thing, but there are some things you probably should not do because you might fall down, like running, skating, playing football, and riding a two-wheeler bicycle. Hold onto the handrail when you walk on the stairs.

Surgery is a trauma to your body. **Surgery to take away FOP bone doesn't work.** The FOP bones just grow back again.

If you have appendicitis or some other sickness that could be dangerous, you may need to have surgery just like anyone else would because the appendicitis could hurt you more than the FOP. That kind of surgery might or might not cause a flare-up. But it wouldn't be a good idea to have surgery you really don't need or surgery to take away FOP bone.

Shots are also trauma and getting a shot can cause a lesion. **People with FOP should not get shots in the muscle.**

These are called **intramuscular** or **IM injections**. DPT (that stands for diphtheria-pertussis-tetanus) shots and flu shots are normally IM injections.

**Some shots can be given under the skin.**

This kind of shot is called a **subcutaneous injection**. It's OK for people with FOP to have

subcutaneous injections. I was given two MMR (that's measles-mumps-rubella) shots under the skin. When people got measles where I live, my doctor thought it would be better for me to get subcutaneous MMR shots than get the measles. I didn't get any lesions from the MMR shots.

**People who have FOP should not have shots of Novocain in their mouths.**

That can make the FOP bone start to grow in your jaw, which could stop your jaw from opening as wide. The dentist can paint on medicine or let you smell a gas that makes you sleepy. These things will stop pain if you need to get a tooth filled and they won't hurt your FOP. If you need to have a blood test or need an IV, the doctor should be careful that the needle doesn't go into the muscle. IV means intravenous and the needle is supposed to go into veins where the blood flows, not into muscles.

**Nobody should hold your arms or legs and try to stretch them.**

You can stretch your own muscles by moving your arms and legs as wide or as high as you can.

But no other person should hold your body and try to stretch it in ways that you can't move yourself.

"Active" stretching where you move your own muscles is good for you. But "passive" stretching if someone tries to stretch your body more than you could stretch it yourself can hurt your FOP.

## Are there things that can help FOP?

A good rule with FOP is to do what feels comfortable. If moving a certain way or sitting in a certain position hurts, your body is telling you that it's not good for you to do it.

**As far as pain, you're the judge; if something hurts, don't do it!**

One thing that can help with FOP is to take good care of your teeth.

-Try not to get any cavities. You should brush and floss your teeth every day. Some people brush after every meal.

-Don't eat sweets, sticky foods, or soda in between meals because the sugar sticks to your teeth and cause cavities. If you do snack on something sweet, brush your teeth as soon as you're done eating.

-There are plaque rinses to keep your teeth clean and fluoride treatments to make them strong.

-Ask your dentist about the best way to take care of your teeth. Be sure to have a check-up twice a year.

Sometimes people with FOP can't open up their mouths. But most of them can still eat food that's cut into small pieces, like meat, cereal, bread, and cooked vegetables. Soft foods like noodles and mashed potatoes work too.

## Should you drink milk?

It's well known that the calcium in milk helps build strong bones. So if you don't drink milk, will FOP bones stop growing? No. It's not calcium that makes FOP bones grow. It's that DNA computer program we talked about. Your body needs calcium for all your bones to grow and stay healthy. You don't need more calcium because you have FOP, but you should drink two or three glasses of milk every day.

Some people with FOP have trouble hearing. It's called a "conductive hearing loss." It won't make you go deaf. Hearing aids should solve the problem.

There are many kinds of tools and equipment that can help you do things. A reacher can help pull things toward you. You can use a gripper to help pick things up from the floor.

Raising a flag or a stick instead of your hand will get your teacher's attention.

You can use the eraser end of a pencil or a wooden dowel to type on a computer keyboard. Utensils like forks and spoons, combs and brushes can have long handles added. Sponges with long handles, a hand-held shower head, and a bench in the shower stall or a bath chair in the tub can help you take a bath or a shower. I have always slept on a waterbed. There is less pressure on your body in a waterbed than on a mattress.

These are just some suggestions to make things easier for you. There are catalog companies that sell all kinds of special equipment and tools. You might invent some things that will help too.

## Can I go to school?

Of course you can. Everybody goes to school.

Because of a law in the United States called "Least Restrictive Environment," kids with FOP should be able to be in regular classrooms at regular schools. The United States government requires schools to be accessible to handicapped people. Public schools have to find ways for all children to learn in school.

I have an aide at school. She's an adult who goes with me all the time. My aide meets the bus when I get to school and takes me to my classroom. She gets out the books I need and sets them up on my desk so I can see them. I turn the pages with a wooden dowel. If my hand is tired I dictate work to my aide. She also stays with me in the halls and the lunchroom to protect me from getting bumped.

I have some adaptive equipment, like a desk with an adjustable slanted top. I put my lunch on a music stand so I can reach it. I use my wheelchair to get around the building. School is one of my favorite places.

## Does anybody else have FOP?

FOP is a very rare disease. It is estimated that FOP affects about 3,300 people worldwide, or approximately 1 out of every 2 million people.

If a football stadium holds 100,000 fans, you would have to fill 20 stadiums before you found one person who has FOP.

That probably means that no one else in your school or even your whole city has FOP.

It may mean that you are the only other person with FOP who goes to your doctor. Your friends and teachers may have never heard of FOP. Sometimes you and your family will have to explain what it is.

That might make you feel like you're pretty much alone.

Not to mention that some of the things you're learning about FOP sound really scary.

But there are other people you can talk to and even meet who have FOP.

You can share your feelings about having FOP with people who know just what it's like. You can ask for ideas about how to make it easier to live with FOP.

## What's the worst thing about having FOP?

The worst thing might be different for each person who has FOP.

- Maybe the worst thing is having a condition that only gets worse, never better.
- Maybe it's having something that's so rare that most people have never heard of it.
- Maybe the stiffness and limited movement is the worst, or maybe it's the pain.

Everybody in the world has some problem sometime in their life. It might be a health problem, or a family problem, or some other kind of problem.

The way for anybody to have a happier life is not to let your problem become so important that you forget all the good things about your life.

## Is there anything good about having FOP?

People can learn and grow from every experience they have. Even though you wouldn't choose to have FOP, your world will be shaped and even strengthened by your experiences with FOP.

Many good things have happened to me because of FOP. I've made friends from all over the United States and the world in the International FOP Association.

My family has gone on vacations to see my friends in the IFOPA. In 1993 we went to an IFOPA reunion in Nashville, Tennessee. In 1994 we went to Disney World.

I've been to Philadelphia for the FOP Symposiums and for appointments with Dr. Kaplan and Dr. Zasloff.



When I was in Philadelphia I learned a lot about the historical things that happened there. In 4th grade I had to do a current events report from an article in a newspaper. Everybody in my class used the local newspaper, but I used the Philadelphia *Inquirer*. That newspaper was founded by Benjamin Franklin! I get special attention because I have FOP, and attention always feels good.

I have some extra services at school, like a special gym class with just 4 other kids. Maybe because I use a wheelchair, everybody recognizes me. Sometimes people know my name when I don't know theirs. Several kids who have FOP have been in the newspaper and on TV.

I'm also pretty good at going to doctors and hospitals. It's not so scary because I go pretty often. And I've learned a lot of words that other kids don't know, like genetic, goniometer, physical therapy, and fibrodysplasia ossificans progressiva.

Having FOP has also made me more understanding of other people who are sick or who have disabilities. I'm not afraid to talk to someone who uses a wheelchair the way some kids might be.

It seems like FOP has made me pretty mature for my age. When I was in the first grade, my teacher said that of all the kids in my class, I had the best self esteem. She meant that I really knew who I was and that I liked myself. Maybe it's the same for you. Kids who have FOP have experiences and things to think about that other kids don't have.

## What can I say when people ask me questions?

You can tell people that you have FOP and that it's a condition where extra bones grow in places they shouldn't grow.

You can say your arm doesn't move because there's extra bone in the way.

If someone says they've never heard of it, you can tell them it's a rare disease. You can say that you were born with it.

Your friends and teachers may have many questions. If they have never heard of FOP, they may want to learn more about it.

**But there may be times when you don't feel like explaining.**

It's OK if you don't want to talk about it. If someone you don't know asks you questions, you can say something short like, "My arms don't move." or "I have a medical condition."

People aren't trying to embarrass you when they ask questions, but if you feel uncomfortable you don't have to talk to them.

## Who else cares about FOP?

There is a support group for people who have FOP called the International FOP Association, or IFOPA.

The IFOPA has approximately 500 FOP members who live in over 55 countries around the world!

The IFOPA publishes a newsletter called the *FOP Connection*.

The *FOP Connection* includes stories about people who have FOP, medical news about FOP, and helpful ideas of ways to make life easier.

Jeannie Peeper started the IFOPA in 1988 because when she was a little girl she didn't know anybody else who had FOP and she wanted to write to other people who had it.

**Her pen pal idea turned into an international organization.**

There are several doctors like Dr. Frederick Kaplan and Dr. Michael Zasloff in Philadelphia and Dr. J. Michael Connor in Glasgow, Scotland who have examined many people who have FOP.

Your doctors can call them to ask questions about FOP.

**In 1992 a laboratory was started in Philadelphia, Pennsylvania.**

Its whole purpose is to understand FOP, find out what causes it, and find a cure for it. Dr. Kaplan

and Dr. Eileen Shore work in the lab along with several other scientists and medical students. They are studying blood, muscles, and tissues from people who have FOP to try to find out what makes their bodies different from people who don't have FOP.

The scientists are making important discoveries every year. All of us in the FOP community hope that soon we will understand what causes FOP and find treatments to stop it.