

What is FOP?

Questions and Answers for the Children



Sarah Steele and Marilyn Hair

*With writings and illustrations by
Children with FOP and their Siblings*

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Children with FOP and their Siblings**

**Recommended for ages 7 to 14
and younger children with adult supervision
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This book was
written by Sarah Steele
and her mother Marilyn Hair.
The “T” voice in this book is
the voice of Sarah Steele.

The 2013,
2nd Revised Edition
of this book was created by Marilyn Hair
and her daughter Sarah Steele
who is 28 years old.

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1

What is FOP?

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

FOP is a disease that causes extra bones to grow in your 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.



2

How did I get FOP?

You were born with FOP. 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 that tells a computer what to do. Well, DNA has instructions that tell your body what to do.

These instructions are called genes. There is a gene that 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. You got FOP because of an accidental 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 and sisters won't have it. Also, FOP is not contagious. Nobody can catch it from you.



3

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. There's even a name for how FOP big toes are shaped. It's "valgus great 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.

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 your 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 have joints that are stuck in different positions. My elbows are closed so I hold my hands in front of my chin. 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 bones 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 the size and shape can change a little so sometimes they seem to get smaller.



4

Does FOP hurt?

FOP 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 when you touch it and it probably hurts.

I feel 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. Pain from a flare-up can last for a couple of weeks or even longer. Then the lesion gets smaller and it usually stops hurting.

It takes anywhere from a few weeks to a few months for FOP bones to grow. When it does, you might feel a hard lump, or maybe you won't be able to move your joint as wide as you could before.

There are a few medicines, but they don't always make a flare-up stop. 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 weather, 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.

5

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 bones that grow in your body don’t go away, and more FOP bones keep growing as you get older.

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, shots or even from having the flu.

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 really hurt and no new FOP bones 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 bones 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!

6

What things could make my FOP get 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 on to the handrail when you walk on the stairs.

Don't have surgery unless you really have to. Surgery is a trauma to your body. Surgery to take away FOP bones 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. That's because the appendicitis could hurt you more than the FOP. That kind of surgery might or might not cause a flare-up.

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

Shots that go into muscle 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. These are called subcutaneous injections. It's OK for people with FOP to have subcutaneous injections. I was given two MMR (measles-mumps-rubella) shots under the skin. I got the MMR shots because kids were getting measles where I live, and my doctor thought it would be worse for me to get measles than to have subcutaneous MMR shots. I didn't get a flare-up from the MMR shots.

People who have FOP should not have shots of Novocaine in their mouth.

Getting a shot in your mouth can make FOP bones start to grow in your jaw, and that can stop your jaw from opening as wide. The dentist can paint on medicine or let you smell a gas that makes you sleepy. That will stop it from hurting if you have to get a filling, and it won't hurt your FOP.

If you have a blood test or IV, the nurse 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 your 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 when someone tries to stretch your body more than you can stretch it yourself is a trauma that can make your FOP worse.



7

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 it hurts when you do something, 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. 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 causes cavities. If you 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, fruit and cooked vegetables. Soft foods like noodles and mashed potatoes work too.

Should you drink milk?

We know 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. I have a piece of plastic tube that's shaped like a cane. I call it my "reacher". I use it to pull things closer so I can reach them. I also use it to scratch itches!

You can use a gripper to 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. Long handles can be taped or screwed onto forks and spoons, and on combs and brushes to make them reach farther.

Using a sponge with a long handle, a hand-held showerhead, and a bench in the shower stall or a bath chair in the tub can help you be more independent in the shower or bathtub.

For a long time I slept on a waterbed because there is less pressure on your body in a waterbed. Now I have a pillow-top mattress. Some people with FOP like to sleep on firm foam like a Tempurpedic mattress.

These are just some ideas that make it easier to do things. There are catalog companies that sell all kinds of special equipment and tools. You might even invent some things that will help!

8

Can I go to school?

Of course you can go to school. Everybody goes to school.

Because of a law in the United States called the Individuals with Disabilities Education Act or IDEA, all kids in the US can go to school in the “least restrictive environment.” Kids with FOP should be able to be in regular classrooms at regular schools. In the US, schools have to be accessible for everybody. 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 my books 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 knocked into by other kids, or falling down.

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

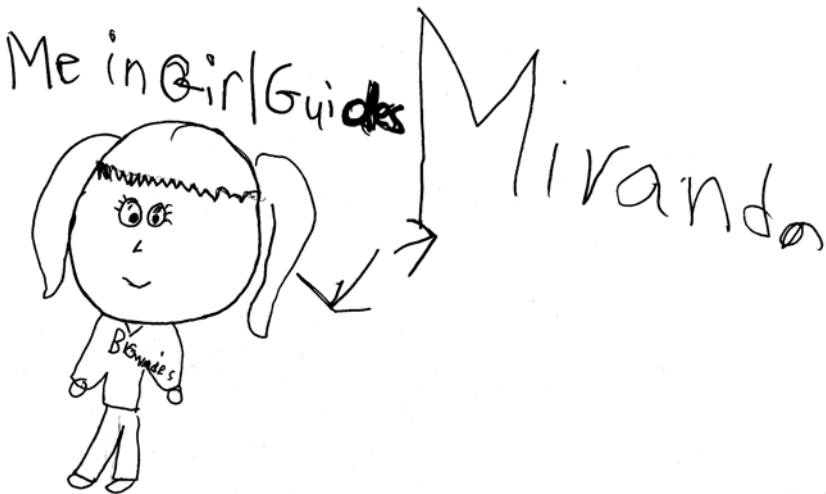
What are some hobbies I might like?

FOP has not stopped people from enjoying many different group activities and hobbies.

Some of these hobbies are reading, sewing, drawing, origami, arts and crafts, collecting things like stamps or stuffed animals, watching TV, going to movies, sending messages to friends and playing games on the computer, traveling, learning a language, listening to and making music, playing in the band or orchestra, and playing cards and board games.

Swimming is a relaxing hobby. Just being in warm water can make you feel better. If you use a whirlpool, the water jets should blow gently so they don't cause trauma to your body.

If you want to ride a bike, there are special bikes that are low to the ground or have more than two wheels to prevent you from falling.



10

What can I do when I grow up?

People with FOP have many different jobs and skills.

Some people who have FOP have their own businesses. One has an auto-parts business. Another does concert promotion.

People with FOP have these careers:

- Banker
- Accountant
- Bookkeeper
- Hotel manager
- Secretary
- Artist
- Teacher
- Newspaper reporter
- News producer
- Actor
- Social worker
- Travel agent
- Mortgage lender
- Lawyer
- Translator
- Tutor
- Computer programmer
- Web designer
- EBay specialist
- Mechanic
- Captain's mate
- Comedian
- Doctor

Every young person has to figure out what they want to do when they grow up. Everybody has to figure out what they like and what they're good at. It's the same when you have FOP. Think about what you like to do, what you're good at, and what will work with your FOP. You can find a way to do anything you want to do.

11

Does anybody else have FOP?

FOP is a very rare disease. Out of 7 billion people in the world, probably 3,500 have FOP. That's 1 out of every 2 million people. The Dallas Cowboys play in a football stadium that has 80,000 seats. You would have to fill the Cowboys' Stadium 25 times 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. You may be the only person with FOP your local doctor has ever met. Your friends and teachers may have never heard of FOP. Sometimes you and your family have to explain what it is.

That can make you feel alone. Not to mention, some of the things you're learning about FOP sound really scary.

But there are other people who have FOP. You can talk to them, meet them and get to be friends. It has been wonderful for me to make friends with kids my own age who have FOP. You can find other kids and adults with FOP in the IFOPA. Maybe you already have!



12

What's the worst thing about having FOP?

The worst thing is probably different for each person who has FOP.

- That FOP only gets worse, it never gets better.
- That most people have never heard of it.
- Stiffness and limited movement.
- Flare-ups and 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.

13

Is there anything good about having FOP?

Nobody would decide to have FOP. But people can learn and grow from every experience. Your world will be shaped and you will even get stronger from your experiences with having FOP.

Many good things have happened to me because of FOP. I have 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. I went to Nashville, Tennessee, Disney World, Florida, San Diego, California, and Philadelphia, Pennsylvania to get together with other people who have FOP. You can find out who is about your age, lives close to you, and has FOP, and get your families together.

Lots of people with FOP go to Philadelphia to see Dr. Kaplan at the FOP lab. There is a lot to see while you're in Philadelphia, like Independence Hall and the Liberty Bell, Betsy Ross's house, the Franklin Institute, the first Ronald MacDonald House, and the University of Pennsylvania.

Several kids who have FOP have been in the newspaper and on TV. One time I was on the local TV news. The reporter wanted to film my everyday life, so the reporter and a cameraman came to my house before school. They filmed me getting up and having breakfast with my family. My little brother told his friends that a TV crew came to his house for breakfast, but his friends didn't believe him.

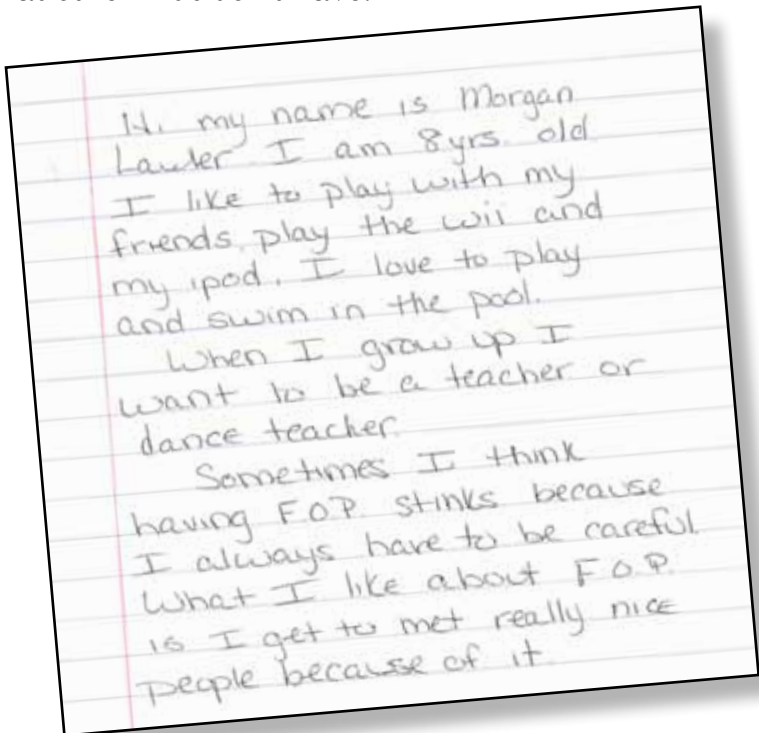
I get special attention because I have FOP, and attention always feels good. I have some extra services at school, like a

special gym class with just 3 other kids. Everybody recognizes me. Sometimes people know my name when I don't know theirs.

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.



14

What can I say when people ask me questions?

When people ask questions, you can say that you have FOP and 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 an 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. You can tell them to search on the Internet and visit **www.ifopa.org**

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.



Fund Research, Find A Cure, Support Families.

My Story

When I first started feeling unwell
And my back and shoulders started to swell
All of my thoughts were scattered
And finding out what was wrong was all that mattered.
Soon we started to see
That everything matched up with FOP.
My family was so sad
Because everything turned out so bad.
When they finally told me
I wanted to flee
But I knew that I had to stay strong
Because home is where I belong
So I bundled up my doubts
And threw them all out
I have no fear
For I know the cure is near

*By: Natalie A. McGuire
Age 11*

15

Who will help?

The International FOP Association helps people with FOP and their families. There are also FOP Associations in other countries. The FOP Laboratory in Philadelphia and other labs at other universities study FOP. Researchers are working to invent treatments and find a cure for FOP.

Here is how the IFOPA began: Long ago in 1988, a young woman with FOP named Jeannie Peeper never knew anybody else who had FOP. Jeannie's doctor was Dr. Zasloff at the National Institutes of Health in Washington D.C. Jeannie asked Dr. Zasloff to tell her about his other FOP patients.

Dr. Zasloff had 18 patients with FOP. My doctor has one patient with FOP – me! Maybe your doctor has one patient with FOP – you!

Anyway, Dr. Zasloff had 18 FOP patients. He gave Jeannie their names and addresses. Jeannie wrote a letter to every one of them, and most of them wrote back. Then Jeannie made a newsletter for people with FOP. She named it FOP Connection. She wanted people with FOP to get to know each other. When more people with FOP found out about Jeannie, she turned her newsletter group into a non-profit organization. It is called the International FOP Association, or IFOPA.

The IFOPA is 25 years old. Now the IFOPA has about 500 members and they live in 50 different countries. IFOPA members speak over 15 different languages! These are some of the countries where people with FOP live: the United States, Canada, China, United Kingdom, Argentina, Brazil, Germany, Italy, France, Spain, Japan, Australia, India, Malaysia, The Netherlands, Poland, Russia, South Africa, Serbia, and Sweden.

Today it's not like when Jeannie Peeper was little and she never knew that anybody else had FOP. The IFOPA still has the FOP Connection newsletter: it's online. And there are lots of ways for people with FOP to get to know each other: The IFOPA has an online directory for FOP members. You can write to people with FOP by letter or email. The IFOPA has an email group called FOPONLINE and a Facebook page. There's also a closed Facebook group that an individual has created for people with FOP. Contact the IFOPA to learn how to access the directory, join FOPONLINE and join the closed Facebook group.

You can get medical papers and books about FOP on the IFOPA website (www.ifopa.org). Also on this site you and your parents can read more about FOP and learn what researchers are discovering about FOP.

There are lots of people who help by giving money to the IFOPA. The IFOPA gives more than \$500,000 every year to the FOP Lab in Philadelphia to help find treatments and a cure for FOP.

The IFOPA sometimes has meetings and family outings. People with FOP can go on vacation to Philadelphia or Disney World. They get to meet each other, learn about FOP, and have fun together.

There are scientists and researchers all around the world who are studying everything about FOP. Twenty researchers, doctors and students work in the FOP lab at the University of Pennsylvania in Philadelphia. Other FOP doctors work together in a group called the International Research Consortium. These doctors work in Australia, Brazil, France, Germany, Korea, the United Kingdom and the United States.

The biggest discovery that was ever made about FOP happened in 2006 in the FOP lab in Philadelphia. The FOP researchers

found the genetic mutation that causes FOP. They discovered the FOP gene! It is the exact tiny change in the DNA that makes FOP.

One child asked Dr. Kaplan, “When you find the FOP gene, what will you do with it?” This is what: The FOP researchers are studying everything about the FOP gene to find out exactly what causes FOP bones. They are always thinking about how to invent treatments to fix the FOP gene to make it stop making FOP bones.

The researchers have made mice that have the FOP gene. The FOP mice make FOP bones, and have babies that have FOP. The researchers are doing experiments with the FOP mice to understand what makes FOP bones grow and find medicine to help the mice and FOP children and adults.





16
P.S.

If you counted 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 other people.

But even though we’re different in some ways, being one in 2 million means we have a lot in common!

The authors hope this book answered some of your questions and has given you ideas about where to find help as you and your family adjust to living with the mysterious condition we call FOP.

Writing and Illustration Credits

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6Hayden Pheif 12 yrs old self portrait
8Sarah Steele 8 yrs old self portrait
12Karina Panasiewicz 5 yrs old
16Miranda Friz 8 yrs old
18-19Morgan Lawler 8 yrs old
26-27Morgan Lawler 8 yrs old
28Arlie Gordon 9 yrs old sister of Zip Gordon

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