

What is FOP?

Fibrodysplasia Ossificans Progressiva
A Guidebook for Families



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This book is dedicated to all of the families who live with FOP every day.

About the cover

The painting on the cover of this book is called “The Circle of Life.” I had a number of reasons for picking this title for my butterfly painting. The butterfly to me is a symbol of hope and new beginnings. It is a subject that everyone can relate to, and everyone has seen a butterfly. Showing the cycle of the monarch butterfly tells of the changes in life which also occur with FOP.

I picked the detailed work of a butterfly in watercolor to show what can be done after my adapting to FOP. I was a right handed painter until two years ago when my right elbow locked, forcing me to now do most of my painting with my left hand. This painting was the first time I had painted an open-winged monarch butterfly using my left hand. I consider this one of the more difficult butterflies to paint. Through my artwork, I also want to show with my painting that people with FOP can have productive lives. It’s important to have a special interest such as painting is to me.

Jack B. Sholund

Bigfork, Minnesota

1995 (for the first edition of *What is FOP? A Guidebook for Families*)

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Foreword

Life doesn't prepare you for fibrodysplasia ossificans progressiva.

Our daughter Miranda Friz was diagnosed with FOP on April 19, 2007. Miranda was 2 years and 3 months old, and a vivacious, fun-loving toddler. Finding out about FOP, and that our daughter has it, was a tremendously painful shock to my husband and me. I have since learned that our reaction was very typical for parents in our situation. For weeks after the diagnosis, we felt our world had imploded. We despaired, and raged, and wondered how this could have possibly happened to our beloved child...

But while we struggled with difficult emotions, Miranda cheerfully carried on —she giggled, chattered, shouted, scattered food on the floor, played with her toys, and did all the things typical for little girls. After some time, Peter and I began to realize that if we wanted Miranda to continue to thrive, we would need to learn how to handle FOP. We couldn't let *it* handle *us*. We started to ask questions: How could we keep Miranda safe? Could she play with her older brother? Ride a bike? Enjoy the outdoors? What about school? There were many issues, and we needed answers.

By then, we were already members of the International FOP Association, and we knew the IFOPA was there to help. As it happens, the IFOPA had a very useful *Guidebook for Families* available on its website. With much trepidation, but even more determination, we printed the guidebook and started to read. Some of the information contained in the guidebook was distressing, and we had to skip over some parts to maintain our equilibrium. For the most part, however, we were grateful and relieved to learn that the guide also contained the vital information we needed. The particular beauty of the guidebook was in recognizing that while there are common challenges faced by everyone with FOP, each family has to determine its own best way to support and protect a loved one with FOP.

The guidebook was an invaluable resource for us. And yet, we couldn't help but notice that it was a little out of date, with multiple references to how difficult it was to research FOP, and how discovering the genetic cause was going to be so important in finding a cure... In fact, by then, the genetic mutation which causes FOP had been known for over a year! Researchers were already making tremendous strides in fulfilling the prophecy of the original guidebook.

Recently, we were pleased to learn that long-time IFOPA member Sharon Kantanie was in the process of revising the guidebook. The version you are about to read is the fruit of

Sharon's efforts as a writer and editor, and is a much-needed update. It is considerably expanded, and contains explanations of the most important advances in scientific knowledge. The guidebook also addresses frequently asked questions about life with FOP, such as whether ear and body piercing is safe, how children with FOP can attend school, and how people with FOP can be immunized.

In addition to practical matters, the new guidebook contains discussions about coping with FOP. I particularly like the section which provides "Families meeting the challenges of FOP." I feel it was a great idea to include a section like this, because for me, it has been enormously helpful to find out how other families have managed with an FOP diagnosis. This section is a good way to begin seeing how people with FOP triumph over challenges, and to introduce you to some fine members of the FOP community.

After an FOP diagnosis, you may feel, as we did, that the earth has been knocked out from under you. Luckily, *What is FOP? A Guidebook for Families* can help you get back on your feet, and help you enable your loved one with FOP not only to live, but live well.

Karen Munro, July 2008

Introduction

Imagine that you are suddenly transported to a new place where everyone speaks a strange new language and nothing seems familiar. You have many questions, yet no answers. Life feels as if it will never be the same. This is often what it is like when parents receive a diagnosis of FOP for their child.

This guidebook is directed to the parents and families of children who have FOP—not because they were the only anticipated audience, but because too often in the past they have been the neglected audience unable to find anything to help them understand what was happening to their child.

The medical topics presented in the first portion of this book try to anticipate questions that families may have or medical situations which may arise. Certain generalizations have been made using the latest clinical and research information in an effort to help parents anticipate the needs of their child. At the same time it is important to note that while many common features of FOP exist in almost everyone who is affected, FOP can affect each person in different ways. Differences among individuals may alter the potential benefits or risks of any medication or treatment option. The final decision about treatment must be made between you and your doctor.

For additional information regarding treatment of flare-ups, please consult “The Medical Management of Fibrodysplasia Ossificans Progressiva: Current Treatment Considerations,” available at www.ifopa.org or by contacting Dr. Frederick Kaplan’s office at the University of Pennsylvania School of Medicine at 215-349-8726. You may also send an e-mail to his assistant at Kamlesh.Rai@uphs.upenn.edu.

FOP is not only a medical diagnosis. It is also a disease which affects many other aspects of life. For this reason, the third edition of *What is FOP? A Guidebook for Families* now features articles on the other challenges that families dealing with FOP face, for example looking at the effects of FOP on family life, promoting independence in children, redefining independence for adults, communicating the special needs of FOP to teachers so that school can be a safe and nurturing environment for learning, and finding resources to help meet the challenges of FOP. Keep in mind when reading these articles that they feature personal reflections of families concerning their unique situations. Readers should talk to their family members and professionals that they trust before making decisions for their own child and family.

The third edition of the FOP guidebook comes at a good time in FOP's history. The gene has been found, associations around the world have been created, and doctors from all around the world have shown an interest in FOP. Hope for a treatment or cure is also stronger than ever.

Note to readers

Some of the chapters that follow reference people with FOP and their families. Ages of individuals are the ages at the time when each piece was written (in some cases as far back as January 2007). Though this book is being published in 2009, a conscious decision was made to do this in order to preserve life and feelings in the present moment.

If you are a parent of a newly diagnosed child, you may wish to process the information in this book gradually. The structure of this book has been set up in such a way so that the information you most need to know comes first—sections like “Just the basics” and “Things to avoid and alternatives.” And if you get no further than the first chapters of the book on your first reading, you may also wish to skip ahead to Chapter 16, “Families meeting the challenges of FOP,” an article in which ten families were asked what they would say to the family of a child newly-diagnosed with FOP, or what they wish someone had said to them. All of us were once where you are now, and somehow we survived. Hopefully you can learn something from our experiences that will make you feel less alone in the face of a rare diagnosis such as FOP.

With special thanks to the following people for their help with this book:

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1.

FOP: Just the basics

Information about fibrodysplasia ossificans progressiva (FOP) can seem overwhelming and filled with new and unfamiliar medical terms. If you are the parent of a newly diagnosed child, FOP can also admittedly seem very scary. So let's start out with some basic information.

General overview

Fibrodysplasia ossificans progressiva (FOP) is a rare genetic condition in which the body makes extra bones in locations where bone should not form. Extra bone develops inside muscles, tendons, ligaments, and other connective tissue. This bone formation outside of the skeleton is commonly referred to as heterotopic ossification by doctors. In people with FOP, bridges of extra bone form across the joints and lead to stiffness, locking, and permanent immobility. In a sense, people with FOP have an extra skeleton.

The condition often begins in the neck and shoulders and progresses along the back, trunk, and limbs of the body. In addition, malformed big toes (short, bent, and sometimes curved inward) are always associated with the condition and can be observed at birth. While the toe malformations cause few problems, they serve as an important early sign of FOP before the onset of extra bone.

Although FOP is congenital, meaning that FOP starts before birth, the extra bone does not form before birth. Symptoms of FOP, including bone formation, usually begin during the first two decades of life. The majority of affected people learn that they have FOP before the age of ten. Inflamed (and sometimes painful) swellings, typically in the shoulder and back areas and sometimes on the scalp or head, are usually the first sign of FOP. The swellings eventually clear up, but they leave behind a new piece of mature bone. People who have FOP experience different rates of new bone formation. In some the progress is rapid, while in others it is more gradual. In each case, the exact rate of progression is unpredictable, although there appears to be a pattern to the progression. For example, extra bone formation tends to occur in the neck, shoulders, and upper back early in life and in the hips and knees during adolescence or early adulthood.

Diagnosis

As previously noted, most people with FOP are born with malformed big toes that are usually short, bent and sometimes curved inward. This is the first sign of FOP, though its significance is typically unrecognized because FOP is such a rare disease. Out of all births, very few infants are born with this specific toe abnormality, but a conclusive diagnosis of FOP cannot be made based on observance of the toes alone. Conclusive diagnosis of FOP is dependent on genetic testing (something that is very new and has only been available since the discovery of the specific genetic change that causes FOP) and observation of other symptoms associated with FOP. The appearance of extra bone will show up on an x-ray. For more information about genetic testing, please consult Chapter 15, “The FOP gene.”

Typically before the age of 10, swellings that look like tumors appear in the neck and back area. Particularly if preceded by trauma, the initial appearance of FOP bone might be in a different area of the body.

At first, the affected areas may be red, painful and sometimes hot to touch. These are common symptoms of inflammation. Some parents note that their children have a low-grade fever in addition to the other symptoms.

Even today, because of the rarity of FOP, many doctors do not correctly diagnose children who have FOP. The diagnostic process sometimes takes months or even years because many doctors are unfamiliar with FOP. The most common explanations that families are given are cancer and fibromatosis (a type of benign tumor). Unfortunately, misdiagnosis and delays in diagnosis can cause great harm, leading to improper tests such as biopsies, which can cause flare-ups and permanent immobility, and inappropriate treatments such as chemotherapy.

History of FOP

FOP or fibrodysplasia ossificans progressiva (*fibro-dis-play-sha os-sih-fih-cans pro-gress-ev-a*) means “soft connective tissue that progressively turns to bone.” The earliest documented cases date back to the 17th and 18th centuries. In 1692, French physician Guy Patin met with a patient who had FOP and mentioned the encounter in his writings. In 1736, British physician John Freke described at length an adolescent whose diagnosis included swellings throughout his back.

The disease became known as myositis ossificans progressiva, which means “muscle turns progressively to bone.” The name was officially modified to fibrodysplasia ossificans progressiva in the 1970s by Dr. Victor McKusick of Johns Hopkins University School of Medicine, who is considered the father of Medical Genetics, in order to acknowledge that

other soft (or fibrous) tissues in addition to muscle (for example tendons and ligaments) are replaced by bone.

How many people have FOP?

It is estimated that FOP affects about 3,300 people worldwide, or approximately one in two million people. Such statistics may be better grasped by the following example: if a large football stadium holds 100,000 fans, one would need to fill nearly 20 football stadiums to find one person who has FOP. At the present time, researchers are aware of approximately 700 people throughout the world who have FOP.

FOP bone and “normal” bone

The extra bone in FOP forms by a progressive transformation of soft tissue into cartilage and bone. This is a process similar to how bone heals after a fracture (break) occurs and is nearly identical to the process by which bone forms normally in an embryo. The abnormality in FOP occurs not in how the bone forms, but rather in the timing and location.

Once it is mature, the extra bone in FOP is indistinguishable from normal bone except by its abnormal location. The bone is strong, can support weight, and will respond to mechanical stress just as normal bone will. In fact, if the extra bone is broken, it will respond just as a normal bone would and heal normally.

How FOP affects muscles

Sometimes an explanation of FOP may be simplified to say that the muscles of people with FOP turn to bone. But this is not really an accurate description of the process that occurs. What we currently know about the process was learned through study of biopsies, observation of skeletons of people with FOP, and additional lab research.

One of the first events to happen during the course of a flare-up is an invasion of lymphocytes and macrophages, special white blood cells that are normally the “good guys” of the immune system, fighting infections and helping us heal. As these white blood cells invade the area, muscle tissue starts to die. Other affected tissue includes tendons (which connect muscle to bone), ligaments (which connect bones to other bones across a joint), fascia (a thin layer that surrounds the muscles) and aponeuroses (connective tissues that join large muscle groups together). As the muscle tissue is destroyed, other cells (special stem cells which have previously been resting quietly) start to divide and multiply and then surround and invade the remaining muscle. Eventually, connective tissue is transformed to cartilage, and then bone. Sometimes the process is quick, and sometimes it takes place gradually. The rate of

bone formation for different flare-ups can be very different depending on which muscle is involved, the stimulus that led to the flare-up occurring, the immune system, and many other factors.

P stands for progressive

Unfortunately, FOP does not improve over time. The “P” in FOP stands for “Progressiva.” That means that FOP will progress, or get worse, as a person ages. As FOP is part of a person’s genetic make-up, people with FOP are born with the condition, even though the extra bone may not have appeared at birth. So people with FOP will not outgrow the condition. Nor can the extra bone that has been produced by FOP disappear.

The body of a person with FOP does not make extra bone all of the time. A person with FOP may go months or years without a flare-up. Yet there is always a chance that extra bone can form, either without any warning (“spontaneous flare-up”) or following trauma, such as a bump, fall, muscle overexertion, an injury, intramuscular injections, surgery, or even some viruses. It is unclear why the disease is active some times and quiet or dormant at other times.

The search for answers—FOP research

FOP research is detective work. The key objectives are to find the right way through a genetic maze to identify the damaged gene that causes FOP (which has now been done) and to use that knowledge to understand what triggers the effects of the disease and develop better treatments and ultimately a cure.

While much of FOP research takes place at the University of Pennsylvania School of Medicine, where a research laboratory devoted to FOP exists, the search for answers is part of a worldwide effort by many individuals and research teams over the past fifteen years. Scientific members of the International Research Consortium in Australia, Brazil, France, Germany, Korea, the United Kingdom, and throughout the United States have identified multigenerational families who helped us discover the FOP gene, as well as making other important contributions to FOP research. People with FOP have generously provided blood and tissue samples and helped to raise the funds that are needed to sustain the research effort. In fact, most of FOP research is funded by FOP families, their friends, and their communities. FOP research is a team effort that could not take place without the efforts of all involved.

2. Things to avoid and alternatives

Let's face it. What you really wanted to do was avoid FOP in the first place. But since that's not possible, please pay very close attention to this information about situations that need to be avoided for people with FOP.

Intramuscular injections and immunizations

Injections into the muscle (intramuscular or IM injections) can be dangerous and can cause a flare-up and bone formation at the injection site. Although intramuscular injections do not always lead to new episodes of bone formation, it is not possible to predict when intramuscular injections will result in a problem and when they won't. As a result, injections into a muscle should always be avoided.

Immunizations and local anesthesia (anesthesia given directly into the area that needs medical help) are two types of shots which are often given into a muscle. A small study of the immunization histories of children with FOP reveals the most accurate information we have on the likelihood of intramuscular injections such as these leading to flare-ups. (Most children have already received their usual childhood immunizations by the time FOP is diagnosed, and as a result have already had some exposure to the "off-limits" injections.) While most individuals seemed to report no complications from intramuscular immunizations, approximately 1/3 of intramuscular injections caused flare-ups within hours of the DPT (diphtheria-pertussis-tetanus) immunization. In fact, in some cases, FOP was first suspected because of a child's adverse reaction to a DPT injection.

Fortunately, there are safe ways to administer most immunizations without needing to resort to an intramuscular injection. Some immunizations, including those that are typically administered into a muscle may be administered under the skin (subcutaneously). Examples of vaccinations that can be administered subcutaneously include measles, mumps, rubella and hemophilus influenza. The polio vaccine can be administered orally (by mouth). Injections and immunizations that are given underneath the skin (subcutaneously) seem to pose little risk. For example, blood tests are obtained routinely without any problems in patients who have FOP. For safest administration of injections, alert the person who will be

administering the shot about the risk of intramuscular injections and trauma. Ask them to use small needles similar to the procedure that has been established for diabetics who need to inject insulin.

Serious consideration should be given to avoiding the DPT immunization, which can only be given into a muscle. Doctors at the Centers for Disease Control feel that subcutaneous injection of DPT might cause serious skin breakdown around the region of the injection. Therefore, it should not be given under the skin. The risk of diphtheria (the D in DPT) is very low. Pertussis (whooping cough) can be treated with antibiotics if it occurs. It is also very rare. Finally, the risk of tetanus is also extremely low unless a child experiences a tetanus-prone injury. If that happens, the hyperimmune globulin could be given intravenously (into a vein) to provide some immunity. The immunization for tetanus can also be given subcutaneously by itself.

A modified subcutaneous administration of the Hepatitis B immunization can also be considered. Normally Hepatitis B immunizations are also intramuscular. There are also vaccinations available for influenza and pneumonia. As with childhood immunizations, these shots are typically administered into muscle. Like many immunizations, they can also be administered subcutaneously (under the skin). See Chapter 11, “FOP and the Flu,” for more information.

In the United States, all states have immunization requirements for children entering school. However, all states offer exemptions to these requirements. The risk of standard childhood immunizations to individuals with FOP should be sufficient grounds for obtaining an exemption. Other countries likely have similar regulations. Please contact your doctor or health system for more information.

Should you have any questions, please contact one of the doctors listed in this book. A list of medical specialists can be found in Chapter 28, “Medical specialists around the world.”

Surgery

Surgery is something that should be avoided unless absolutely necessary. For people with FOP the surgery that is most often considered (or at least talked about) is surgery to remove the extra bone that gets created by FOP. But surgical intervention to remove extra bone or attempts to try to improve a person’s condition surgically often results in the exact opposite: more robust bone growth and a worsening of the condition. New bone will almost certainly grow back and further impair mobility. As a more complete understanding of the genetic and molecular basis of FOP evolves, perhaps safe surgical removal of the extra bone will

eventually be possible. At the present time, however, this type of surgery should be avoided because of the great risks involved. In addition to the high risk of additional flare-ups, there is a high risk of complications such as infection and phlebitis (an inflammation of a vein). This is especially true with surgery of the lower limbs. It is also important to note that such surgical intervention often ends in failure, as it is difficult to reposition one set of joints in the lower limbs without affecting posture and balance.

While surgical operations usually make FOP worse by causing the body to generate more bone, emergency situations may exist where an operation may be necessary, as in appendicitis or acute gall bladder disease. Although such an operation may lead to a flare-up, the life-threatening nature of the emergency may justify the surgery.

The following guidelines may be helpful in dealing with surgery and FOP. Some suggestions involve advance planning so that you can be prepared in case an emergency happens.

- Be aware of the risks of new bone formation following injury to the musculoskeletal system or surgery.
- Avoid elective surgery on the musculoskeletal system. Although the bone can be removed, it will often grow back and lead to problems that are generally worse than the original condition. Surgical procedures to increase mobility do not work.
- Ask your family physician or internist to schedule a consultation for you with a lung specialist and perhaps an anesthesiologist in your local area who can work out a safe plan for administering a general anesthetic should an emergency arise. As people with FOP have unique respiratory issues and/or fused jaws, you may want to consider doing this in advance of an emergency so that a plan is in place should the need arise. Such a plan could be filed in your chart, and a copy should be made available to you, particularly when you are out of town. You may also wish to consult Zvi Grunwald, M.D. an anesthesiologist who has treated many cases of FOP, for additional information. He can be reached at 215-955-6161 or zvi.grunwald@jefferson.edu.
- If emergency surgery is needed, careful planning and management of airway problems should be recognized and implemented.
- Avoid intramuscular injections.
- It is important to note that in all medical situations one should be prepared to educate doctors, emergency medical technicians (EMTs), and hospital personnel about caring for someone with FOP. Consider organizing personalized medical information about FOP and other health needs in advance. Having personalized medical information at your fingertips helps reduce stress and in some cases provides the opportunity for better care. To obtain a “Medical World Binder,” an

organizational system for medical documents, contact the IFOPA office at together@ifopa.org or call 407-365-4194.

Read Chapter 7, “Emergencies,” for more information on surgery and FOP, as well as for additional suggestions for communicating with medical professionals about the unique issues of FOP.

Falls

No one tries to fall. Falls happen. In childhood, they may happen during play. During adulthood, they may happen as the extra bone interferes with balance. Unfortunately, they pose a serious risk to people with FOP. Falls can lead to flare-ups, or in very severe cases, head injury, loss of consciousness, concussion, neck or back injury and even death. A study showed that people with FOP are twice as likely to suffer serious effects from falls as compared to individuals who do not have FOP.

The ability to keep one’s balance is required for stable gait (walking ability). Unfortunately, there are numerous factors which reduce this ability in people with FOP. Because of the diminished flexibility of the neck and chest wall, visual input is reduced to a limited field of view. Even if sensory input alerts the patient to a dangerous situation, motor response is limited due to joint fusion and muscle involvement. Mobility restriction from fusion of the neck, trunk, and limbs also severely impairs the balancing mechanisms and protective responses to falls.

There are, however, things that can be done to make an environment safer and lower the risk of falls:

- Remove things that a person can trip over (papers, books, clothes, shoes, etc.) from the floor.
- Use non-slip waxes on wood floors.
- Remove or limit the use of rugs. Use double-sided tape to keep rugs from slipping.
- Install grab bars next to the toilet and in the tub or shower.
- Use non-slip mats in the bathroom. Also consider using one inch unglazed ceramic tiles, as they make it less likely that a person will fall on a wet floor.
- Improve the lighting in your home. Consider installing nightlights to add extra light at night. Some lights can even turn on by themselves when it gets dark.
- Install handrails and lights on all staircases.

- Remove caster wheels from furniture. Remove any furniture that is unstable when leaned on.
- Make sure that power cords are not in the walking path.
- Have your (or your child's) vision and hearing checked regularly.
- Be aware if any medications you (or your child) are taking can cause drowsiness or interfere with balance.
- Watch out for pets!
- Use shoes that fit well and provide good balance.
- Use walking aids (crutches, canes, walkers, etc.)

Fear

In some ways, this may be the hardest thing on the list to avoid. The uncertainty of FOP can sometimes cause you to be fearful of what might happen. You've quickly learned that bumps, falls, intramuscular injections, and surgeries that would not be a problem for someone else can lead to an FOP flare-up, and you've also learned that FOP strikes mysteriously for no reason at all. Particularly when FOP is new, it can be hard not to be scared of flare-ups and to wonder whether there is something that you could have done to change the situation so that a particular flare-up didn't happen. Our own emotions can be our biggest enemies sometimes. Just take one day at a time and try not to be too hard on yourself. Although FOP will change the way that you and your family live, you will learn to adjust to a new way of life with determination, strength and courage.

3.

Flare-ups

A flare-up is the common name for the symptoms of active FOP. The topics that follow are designed to help you understand more about the unique symptoms of FOP, as well as how to manage them.

What causes a flare-up?

The simple answer is that we don't really know what underlying process leads to a flare-up. A flare-up can start as a result of trauma (a bump, fall, injury, overexertion of muscles, intramuscular injection, surgery, etc.). There are also times when, in spite of obvious trauma, a flare-up does not occur. Many times flare-ups will also start with no apparent reason. Based on evidence that suggests a link between influenza (and perhaps other viruses) and flare-ups, it is possible that at least one trigger of FOP flare-ups may be based in the immunological system. This explanation would make sense, as swelling and inflammation are immune system reactions. But at the present time the immunological features of FOP are still poorly understood.

Forming bone

A flare-up occurs when the body starts to generate new bone, although not every flare-up results in a completion of the process. No one knows what initiates this process, but once it begins, inflammation, tissue swelling, and discomfort follow. While flare-ups are usually painful, the degree of pain can vary. In addition, sometimes the individual will not feel well and may develop a low-grade fever.

A single flare-up may continue for as long as 6-8 weeks, or sometimes even longer. Multiple flare-ups may also occur during an active period of FOP. As stated earlier, a flare-up's length often depends on which muscle is involved, the stimulus that led to the flare-up, the immune system, and many other factors that we still don't entirely understand.

Presently, there is no medication or therapy that can stop the process of bone formation once it has begun. However, a physician can prescribe medicine that can help minimize the flare-up (so that hopefully less bone will form) and help relieve the inflammation and pain. Medication should be started as soon as possible after symptoms appear. Refer your physician to "The Medical Management of Fibrodysplasia Ossificans Progressiva: Current

Treatment Considerations” (posted on the IFOPA website or available through the University of Pennsylvania School of Medicine).

When a flare-up is in progress, joint stiffness may occur overnight. One should not think that a piece of bone grew overnight—it takes anywhere from several weeks to several months for bone to form. The stiffness comes from swelling and pressure inside the muscle during the earliest stages of new bone formation.

Many people have noted that flare-ups in adults can be different in character from those that occur in children. It appears that children tend to have more nodular flare-ups (lumps), while adults tend to have flare-ups that involve swelling of the whole limb. Although these differences have been noted, scientists do not yet understand the reason for these different patterns may occur. Although one type of flare-up may be more common than another at a particular age, either type can occur in anyone who has FOP.

Flare-up symptoms can also vary depending on which muscles and joints are affected. For example, flare-ups in the hip area are less likely to produce lumps or obvious swelling, as the hip muscles are very deep—compared to other muscles that are closer to the surface of the body. Disabling hip flare-ups may begin with nothing more than a sensation of a strained hip or groin muscle.

Normally in flare-ups there is a progressive transformation of soft tissue into cartilage and then to bone. However, it should also be noted that some flare-ups seem to stop at the cartilage stage. We know this with certainty because physical examinations will sometimes reveal a firm bar, sheet, or plate of tissue that does not show up on an x-ray. Mature bone will appear in x-rays, however cartilage will not. Cartilage may be a term you have heard before, but exactly what is it? Cartilage is a type of connective tissue that serves to provide structure and support to the body’s other tissues without being as hard or rigid as bone. It can also provide a cushioning effect in joints. Of course, like the extra bone created by FOP, the cartilage that forms as a result of some flare-ups is in places where it shouldn’t be. We currently do not know why some flare-ups seem to stop at this stage.

Lumps and bumps

Soft lumps often appear for no apparent reason on the neck and back as early as the first year of life. The lumps may be very small or quite large and may appear overnight. They are signs of swelling and/or inflammation. Occasionally the lumps disappear, but more commonly they mature to form a new piece of bone. Although the bone has all of the characteristics of the normal bone in our skeletons—complete with marrow—FOP bone forms in places

where it should not form, such as in muscles, tendons, and ligaments. The process that occurs completely replaces these structures with bone.

The lumps initially are soft, often painful, and may be warm to touch. After they turn into bone, they often stop hurting, although they still may be a source of discomfort due to pressure over those areas. Once the lumps turn into bone, they become part of a person's body. They occasionally may seem to change their shape and size, much like the bony bump on the outside of a broken bone.

When the soft lumps first appear, they may not be recognized and diagnosed as FOP and may be mistaken for tumors or cancer. Often the lumps are biopsied and misdiagnosed. Frequently, the surgical trauma of the biopsy leads to additional bone formation at that site or in nearby joints (for example shoulders fusing after a biopsy under the arm).

Sometimes the first symptom of FOP is not a typical flare-up, but rather a swelling or bump in the scalp portion of the head. This type of swelling can be present as early as the first month of life. These swellings affect a type of connective tissue known as aponeuroses, a thin layer of tissue that separates muscles from each other. It is important to know that this type of swelling does not affect the brain at all and does not seem to cause any problems, no matter how much swelling occurs.

FOP and pain

Some people find that most or all of the pain subsides when a flare-up is over. It appears that it is the process of extra bone formation, rather than the extra bone itself, that usually leads to pain. FOP is not always painful. However, particularly in more advanced cases of FOP, pain can be more chronic in nature. This may be because the extra bone applies pressure to the nerves and/or muscles. Chronic pain may also occur as additional bone develops and the already existing bone makes it harder for the body to compensate for new restrictions in mobility. Again, consultation with a physician is recommended to help manage these symptoms.

It's also important to remember that a person with FOP can still have normal aches and pains. The best rule with FOP is to do what feels comfortable. Sometimes a hurt or ache is the body's way of telling us that we are doing too much. In FOP, it is a good idea to avoid those activities or positions that cause discomfort. When muscles are stretched, they often react by pulling back in the opposite direction, creating tension in the body. In some ways, it is like stretching a rubber band; the more it is stretched, the more it pulls back. This causes

additional strain on the body. The key with FOP is to avoid activities that cause pain or are likely to lead to injury.

In many cases, you will know if an ache or pain is caused by a flare-up because the pain will be accompanied by other FOP symptoms such as swelling and inflammation. One notable exception to this may be hip pain. The muscles in the hip are deeper muscles, so there may not be visible swelling or inflammation. If there is a doubt as to whether the pain is related to a flare-up or some other cause, you may wish to consult your physician.

A common medical test is an x-ray. An x-ray will show bone, but not in the early stages of bone growth. X-rays are not very helpful in determining whether pain is a symptom of a flare-up.

X-rays involve radiation. Please only perform this test when your doctor feels it is necessary. If you have additional questions about when this test may be useful, please consult one of the medical professionals listed in Chapter 28, “Medical specialists around the world.”

For more information on managing pain, see Chapter 8, “Treating acute and chronic pain.”

4.

FOP and mobility

In individuals with FOP, bridges of extra bone form across the joints and lead to stiffness, locking, and permanent immobility. Learn more about how FOP affects mobility and how to stay as mobile as possible.

How FOP affects mobility

FOP can affect all of the regions of the body as it lays down what is essentially an extra skeleton. The progression of extra bone growth, medically known as ossification, follows a characteristic pattern. Usually flare-ups and extra bone progress from head to toe, from back to front, and from upper limbs to lower limbs. This means that FOP typically starts in the neck, spine, and shoulders before developing in the elbows, hips, and knees.

The muscles of the diaphragm, tongue, eyes and heart are not directly affected by FOP, meaning that extra bone does not seem to form in these muscles. However, it is important to note that while FOP spares internal organs, crowding of organs can occur as a result of the extra bone growth. The well-documented and characteristic progression of FOP, as well as the regions not affected, likely holds important clues to the cause and development of the disease.

The body's joints, such as the knees or the elbows, connect the bones and aid in movement. In FOP, extra bone replaces and crosses the ligaments (which cover the joints), as well as muscles and tendons (which move the joints). Consequently, movement of joints in areas affected by FOP may become difficult or impossible.

Different people are affected differently

Most people who have FOP have similar features, particularly malformation of the great (big) toes, which is present at birth, and extra bone formation which progresses throughout life. However, much variation exists from person to person. The largest variation occurs in the timing and rate of extra bone formation. For example, one person with FOP may lose motion in the hip during the first decade of life, while another person may still have normal walking mobility into adulthood. Another common variation includes the severity of disability. For example, an elbow could be fixed in a bent position (putting the arm

permanently across the chest), fixed in a straight position, or may be left with some range of motion.

Can anything be done to help a person with FOP maintain mobility?

Prompt treatment with medications may help minimize a flare-up, but once the process of building extra bone starts, there is sadly little that can be done to interrupt the process. Again, it's the extra bone inside and crossing muscles and joints that takes away mobility. While a person with a typical injury can do physical therapy to try to regain mobility and/or strength, physical therapy is not recommended for individuals with FOP. This is because physical therapy generally involves stretching of the muscles, sometimes passively where the therapist does all of the work, and sometimes with active help from the patient. Even mild stretching can lead to a new flare-up or worsen an existing one. Instead, it is best to focus on movements that are comfortable and part of every day living. Staying as mobile as possible within the limitations of FOP is the best way to keep up muscle mobility and strength. Swimming, warm water hydrotherapy, or being in water are also good activities, and fun too. The unique properties of water reduce the pressure on muscles and may make it easier to move.

Physical activities

Because of the risk of trauma, parents should consider having their child with FOP avoid physical activities with high likelihood of getting hurt or falling (running, contact sports, etc.). As mentioned previously, people who have FOP have an increased tendency to fall due to impairment of balance from stiff joints, and if a person who has FOP begins to fall, it is less likely that he or she will be able to break the fall. There is also some anecdotal evidence that overexerting the muscles can contribute to flare-ups.

However, given FOP's progressive nature, a parent should let a child take advantage of the things that he or she physically is able to do. It is important not to isolate children from the joys of life or the socialization with friends. In other words, don't be so afraid that your child will get hurt that you and your child forget that trying new things and having fun doing so is an important part of life. Life is sometimes about balancing risks.

Your family can make modifications to conventional games which make them safer and/or easier to play. To pursue adaptive sports or get help in making modifications to sports and activities, contact the following organizations:

- Cure Our Children Foundation (website with detailed information about sports for people with disabilities, includes information about specific sports and international resources); 310-355-6046, www.cureourchildren.org/sports.htm; website includes the article “I Know I Can Do It: Sports Are For Disabled Children Too”
- Disabled Sports USA, Far West; 530-581-4161, www.dsusafw.org
- America’s Athletes with Disabilities; 800-238-7632, www.americasathletes.org
- American Association of Adapted Sports Programs; 404-294-0070, www.adaptedsports.org
- National Center of Physical Disability and Activity; 800-900-8086, www.ncpad.org
- Children’s Golf Foundation; 561-842-0066, www.childrensgolf.org
- Courage Center; 888-846-8253, www.courage.org
- Adapted Physical Education (site with suggestions for adapting PE activities); 540-953-1043, www.pecentral.org/adapted/adaptedmenu.html
- Kids Camps (resource for finding camps for children with disabilities); 877-242-9330, www.kidscamps.com/special_needs/physical_disability.html
- Most of these resources are US-based. For information on adaptive sports in other countries, refer to Cure Our Children Foundation, physicians, schools, recreation centers, etc.

Getting around

As FOP progressively restricts the ability to walk, a person who has FOP may find canes, crutches, walkers helpful in maintaining walking mobility. Particularly for distance walking, many people whose legs are affected by FOP find that it is easier to get around in a power wheelchair or a motorized scooter.

Advantages to wheelchairs and scooters include the following:

- Safety. As FOP restricts movement and balancing becomes more difficult, wheelchairs and scooters are safer alternatives for getting you where you want to go safely and quickly. It also helps protect a person from getting bumped by people in public places, who can sometimes be absent-minded or distracted so that they don’t look where they are going.
- Speed. Power chairs are a fast way to get from one place to another.
- Comfort. A wheelchair can be custom made especially for a person’s needs, from special seating to a sit-to-stand chair that will take a person from a seated

- position to a standing one. A wheelchair can also help a person get around without getting tired.
- Independence. A lot of people think a person becomes more dependent when using a wheelchair. But with a power chair, a person becomes more independent. It's easier to move on your own and perhaps even be a little more adventurous.

People with FOP often need customized wheelchairs, particularly when limitations progress to the point when a power wheelchair is required for preservation of independence. A good place to turn for assistance is a seating and wheelchair clinic at a rehabilitation center or hospital. Wheelchair clinics have teams of specialists including physiatrists (rehabilitation doctors), occupational and physical therapists, nurses, and wheelchair vendors. These teams are typically familiar with the wide variety of wheelchair options that are available, from wheelchairs that can recline and/or elevate to those that can take a person from a seated position to standing—and in some cases even drive from a standing position if needed.* They can help customize seating to maximize comfort and make sure that the wheelchair is suitable for home or school use.

**The Swedish wheelchair manufacturer Permobil's "Stander" model has all of these features (though its footplates are a few inches off the ground), and other Permobil chairs can also be adjusted to have anterior tilt, which may create easier/more independent entry from a standing position. Permobil wheelchairs are currently available in the United States, many European countries, Canada, Japan, and Korea. Other manufacturers also make wheelchairs with standing features. The Permobil is being highlighted because it has frequently been chosen by people with FOP. Contact FOP associations or rehabilitation centers in your country to learn about power wheelchairs with similar features.*

5.

The medical world

This chapter can help you better communicate with medical professionals.

Introduction

Rare disorders present a unique challenge in the medical world. On the one hand, families are searching for answers and treatments. Doctors and medical professionals, on the other hand, have many patients with more common problems and cannot be experts on every rare disorder. The purpose of this chapter is to help families navigate the health system to receive the best possible care.

Selecting a doctor

Since most doctors have likely never heard of FOP, it is not essential to find one who has to be your primary physician. The best doctor for a person with FOP is one that will listen to you, be open to learning about FOP and potential treatments, understands the issues involved with treating someone with FOP, and is willing to consult with Dr. Kaplan or any of the other medical professionals listed in chapter 28 when needed. Your primary physician may be a pediatrician, a family doctor, an internist, an orthopaedist, or a rheumatologist. Again, it is mostly about who you feel most comfortable working with to meet your needs and deal with any health concerns that arise. FOP.

Helping your doctor provide better care

It is extremely important that any relationship with a doctor be a partnership. People with rare disorders and their parents often become quite knowledgeable. After all, parents know their child best and are familiar with day-to-day care and even at a young age people with FOP develop self-awareness about their needs.

Share as much information as you can with your doctor. Consider providing him or her with the following information:

- “The Medical Management of Fibrodysplasia Ossificans Progressiva: Current Treatment Considerations,” available at www.ifopa.org or by contacting Dr.

Frederick Kaplan or his assistant Kay Rai at the University of Pennsylvania School of Medicine at 215-349-8726 or Kamlesh.Rai@uphs.upenn.edu.

- A copy of *What is FOP? A Guidebook for Families*
- The FOP emergency card (see below for more information)
- A personalized “Medical World Binder” that contains information on your child’s (or your) medical history
- Consider having a one page summary sheet of personal medical information
- When updates become available, let your healthcare providers know so that they always have the latest information.

Some of these resources are available through the IFOPA, whose contact information is as follows:

IFOPA

P.O. Box 196217

Winter Springs, FL 32719-6217

Telephone: 407-365-4194

E-mail: together@ifopa.org

Website: www.ifopa.org

When you see your doctor, come prepared with a list of questions so that you can use your time most effectively. Don’t save your most important question for last so that your doctor realizes its importance and it can get the time and attention it deserves.

Ways to share medical information

The IFOPA has developed the following ways to educate medical professionals about FOP. First, the FOP emergency card is quick source of vital medical information. It contains the following:

- Includes basic information about FOP
- Emphasizes that deep tissue trauma accelerates the condition and persons with FOP must be handled gently
- Lists emergency precautions that should be taken fore treating a person with FOP
- Explains that intramuscular injections should be avoided because they can cause a flare-up
- Provides emergency contact information for Drs. Kaplan and Pignolo
- Provides information about the urgent need for tissue collection from a person with FOP in an emergency situation

The FOP emergency card can be used in the following ways:

- Carry one in your wallet
- Give one to your caregiver
- Loan one to a babysitter
- Leave one in the family car
- Put one in a pouch to be worn by your child during a field trip or play date
- Give one to the school and/or camp nurse

Consider supplementing the card with contact information for local doctors and your own emergency contact information. Cards can be obtained at no cost from the IFOPA.

Another helpful resource is the “Medical World Binder” of personalized medical information. The IFOPA provides a kit, which includes the binder and the following items:

- Identification sheet
- Emergency contact phone number sheet
- Business card holder that can be used for cards from medical professionals whom you encounter. Consider also including extra FOP emergency cards there.
- Prescription holder
- Personal medical history information
- Tabs – For creating sections in your binder. Suggested sections are 1. Tracking flare-ups, medications, and supplements 2. Hospitalizations 3. Medical appointment notes 4. Tracking system for medications
- CD holder – for CDs of medical tests such as MRIs
- Plastic document holder for duplicates of important documents

Contact the IFOPA for more information on this kit.

The IFOPA also recommends that each family participate in Medic Alert. Medic Alert is a non-profit membership organization that was founded decades ago to provide continuous access to medical information in an emergency. The member wears a symbol on a necklace or bracelet that is recognized by emergency responders around the world. When they see that logo with your personalized identification number and medical condition, they know to call the Medic Alert 24-hour emergency response center immediately—even before treatment—unless the life of the patient is at risk from a few moments delay.

Because of FOP's rarity, just putting the name of the condition on the bracelet or necklace will not communicate any useful medical information. Therefore, after consultation with firefighters, emergency medical service attendants, physicians, and Dr. Kaplan, we suggest the following working for the Medic Alert information: "Excessive bone formation due to genetic disorder. Must handle gently."

In order to join Medic Alert, visit www.medicalert.org or call 800-432-5378 or 888-633-4298. For people calling outside the U.S. (including collect calls), please dial the following: 209-668-3333. They have a language line to accommodate international families. The fee varies depending on which form of identification you choose and includes the identification bracelet/necklace, laminated identification card, unlimited updating of your records, and accurate medical data 24 hours a day.

6.

Making FOP treatment decisions

The following section is an introduction to treating FOP. Although there are common physical features shared by every person who has FOP, there are differences among individuals that may alter the potential benefits or risks of any medication or treatment discussed in this chapter. The final decision about treatment should always be between you and your doctor. Please also consult “The Medical Management of FOP: Current Treatment Considerations,” available on the IFOPA website at www.ifopa.org or through the University of Pennsylvania School of Medicine. Contact Dr. Frederick Kaplan’s assistant Kay Rai at 215-349-8726 or by e-mail at Kamlesh.Rai@uphs.upenn.edu.

Introduction

While there is no proven effective prevention or treatment for FOP, a better understanding of the underlying cause of FOP is leading to new medication-based strategies to treat FOP. More than any time in the past, physicians are faced with an increasing number of potential medical interventions.

Unfortunately, the rarity of FOP and the unpredictable nature of the condition make it extremely difficult to assess any therapeutic intervention. Because FOP symptoms can come and go, it can sometimes be hard to tell if a particular treatment has truly been successful or if a flare-up has just “run its course.” Add to that our own wishful thinking that there is something that we can do to affect the outcome of a flare-up, and you can see why it is so difficult to have a truly objective discussion of FOP treatment options.

This report reflects the experience and opinions of the University of Pennsylvania research team and the International Clinical Consortium on FOP on medications which have been used in the treatment of FOP. This information is meant only as a guide. Again, although there are common physical features shared by every person who has FOP, there are differences among individuals that may alter the potential benefits or risks of any medication or treatment option. The final decision about treatment must be made between you and your doctor.

Yes, it's a bit confusing

At the outset of this section of the book, let's set the record straight. Despite best intentions to keep medical explanations as simple as possible while still providing complete information so that families can have the facts needed to make wise decisions about medical care, this section gets more technical than most other sections of the book in some areas because of the nature of the subject matter. Please do not let this frighten you away from reading it! Instead, try to understand what you can. If you encounter something that seems a bit confusing, please ask questions of one of the "FOP medical specialists around the world" or ask your own doctor. The medical system works best when doctors and patients work as a team. It is important for you to understand and consider the FOP treatment possibilities so that when a flare-up does happen, you and your child's doctor will already have a plan in place regarding what to do.

Introduction to corticosteroids (prednisone) and anti-inflammatory medications

There are several general classes of medications that are currently used to treat FOP flare-ups. The first group includes medications that have been widely used to control the symptoms of flare-ups (swelling, inflammation and pain) and have had positive anecdotal reports from families with generally minimal side effects. Examples of these medications include short-term use of high-dose corticosteroids (such as prednisone) and use of non-steroidal anti-inflammatory drugs (NSAIDs) including the new anti-inflammatory Cox-2 inhibitors, more commonly known by their brand names such as Celebrex (a common arthritis drug), Celebra, or other names depending on your location.

Prednisone

Prednisone has potent anti-inflammatory effects, and for this reason it is often considered for treatment of FOP flare-ups. For maximum beneficial effect, prednisone should be started within 24 hours of the onset of a flare-up. The medication is generally less effective if started when a flare-up is more than two days old. Therefore, it is extremely important to contact a physician as soon as you notice a flare-up starting. Some families find it helpful to have a supply of prednisone at home in case of an emergency. If the flare-up responds to prednisone but comes back when the medication is discontinued, you may consider repeating a four day course with a 10 day taper.

Prednisone should not be used for flare-ups on the chest or trunk, as it is difficult to judge the exact onset of a new flare-up. On the other hand, flare-ups of the jaw can become medical emergencies which interfere with breathing and eating, so prompt use of prednisone

is especially important. This potentially dangerous type of flare-up may also require a slightly longer course of treatment with a gradually tapered dose until the swelling subsides. Remember that this is an exception to the usual recommended treatment.

Use of prednisone may also be considered after soft tissue injury in order to try to prevent the start of a flare-up and to minimize effects of the injury. It should not be used after minor bumps and falls.

Prolonged or chronic use of corticosteroids such as prednisone is of no benefit. In fact, there is some evidence that long-term use can actually accelerate bone growth and harm the body in other ways (for example by suppressing immunity, affecting vision, contributing to osteoporosis, causing adrenal insufficiency, etc.). The use of prednisone for FOP treatment is only meant to suppress swelling, inflammation and hopefully abort the early migration of lymphocytes (those special white blood cells we talked about earlier in this book) into the muscle and potentially keep the muscle from dying and bone from growing in its place. See “The Medical Management of FOP: Current Treatment Considerations” for more information, including recommended dosages.

Alphabet soup—Cox-2 inhibitors and NSAIDs (Non-Steroidal Anti-Inflammatory Drugs)

When alternatives to prednisone are desired, when prednisone use is discontinued, when a flare-up is more than 48 hours old at the start of treatment, or when long-term treatment is needed, Cox-2 inhibitors or non-steroidal anti-inflammatory drugs (NSAIDs such as ibuprofen, naproxen, etc.) are another option.

These medications specifically target special inflammation-causing substances called prostaglandins. These prostaglandins also support new bone formation. Studies in the medical literature have shown that lowering prostaglandin levels in experimental animals dramatically raises the threshold for heterotopic ossification, making it more difficult for extra bone to form.

In addition to their potent anti-inflammatory properties, a recent study unexpectedly demonstrated that the new Cox-2 inhibitors have potent anti-angiogenic properties, a feature that makes them even more desirable for consideration in FOP. What this means is that these medications help prevent certain new blood vessels from growing. These blood vessels help “feed” the growth of extra bone. So if we can stop or slow down the growth of this system of blood vessels, then perhaps we can stop the extra bone growth.

However, the data suggest that in order for prostaglandin inhibitors to be truly effective in preventing extra bone growth (heterotopic ossification), the medication must be “in the system” (in other words circulating in the blood at therapeutic levels) before a bone-producing signal occurs.

You are probably more familiar with the brand names of Cox-2 inhibitors, which are often used to treat arthritis and other conditions that cause inflammation. Brand names may include Celebrex, Celebra or other names depending on where you live. Compared to NSAIDs, which have been around longer, Cox-2 inhibitors offer the possibility of fewer gastrointestinal side effects/risks. Also, they only need to be taken once or twice a day. But recently substantial concerns have been raised about the safety of Cox-2 inhibitors, including a recall of several drugs in this class after an increased risk of heart attack and stroke was discovered in patients who were part of research studies involving the recalled medications (Vioxx, Bextra). While the gastrointestinal benefit of Cox-2 inhibitors over traditional non-steroidal anti-inflammatory medications remains in question, Cox-2 inhibitors seem to be a reasonable choice for people with low cardiovascular risk (low risk of heart problems) who have experienced serious gastrointestinal difficulties in the past or people who are at high risk of gastrointestinal problems, such as people with FOP who may need to use corticosteroids (prednisone) from time to time (intermittently) or at the same time (intercurrently) as other anti-inflammatory medications.

With all anti-inflammatory medications, careful gastrointestinal precautions should be taken. If long-term use of a Cox-2 inhibitor is considered, certain blood tests should be administered to periodically monitor serum liver and kidney function. Because of the risk of heart attacks and/or blood clots, Cox-2 inhibitors should be used with caution in people with a history of heart disease, diabetes, high cholesterol, or in people with significant immobility that leaves them unable to walk (which puts them at a higher risk for blood clots).

See “The Medical Management of FOP: Current Treatment Considerations” for more information, including recommended dosages.

What are aminobiphosphonates and why could they be helpful in the treatment of FOP? (Pamidronate and Zoledronate)

Aminobiphosphonates are a class of medication which act primarily to inhibit bone resorption, or stop bone loss. Two drugs in this class are Pamidronate and the more potent

zoledronic acid (Zoledronate/Zometa). Currently more information is available pertaining to the use of Pamidronate and its potential use in FOP treatment because Zoledronate should not be used on anyone younger than 18 years of age.

At first, there seems to be little reason for using aminobiphosphonates in the treatment of FOP since our desire is to stop bone growth from occurring. However, the story is not that simple.

All medications have side effects, but it is an interesting part of medical practice that, on occasion, medications have been used either mistakenly or coincidentally with unanticipated beneficial effects. Such a scenario occurred in recent years with the use of the aminobisphosphonates in the treatment of FOP. Several anecdotal reports (to Drs. Kaplan and Glaser at the University of Pennsylvania) from physicians and FOP patients highlighted the response of FOP flare-ups to Pamidronate, one of the newer aminobisphosphonates. But why would Pamidronate even be considered for the treatment of FOP flare-ups? Ironically, in all three cases reported to the University of Pennsylvania research team, the medication had been used with the mistaken belief that Pamidronate was more potent than Etidronate (a medication/biphosphonate previously used in treating FOP without success) in inhibiting mineralization, a process that leads to bone formation. It is not. None of the newer bisphosphonates, including Pamidronate, have any effect on suppressing mineralization. Nevertheless, all three patients and their physicians independently reported substantially decreased swelling, redness, and pain following high dose intravenous Pamidronate administration during a new flare-up. In one patient the Pamidronate was administered alone, while in the other two patients, it was administered along with an oral steroid (such as Prednisone) for several days during the early phases of a new FOP flare-up.

At the time of printing Pamidronate use has been reported in 13 patients. In 10 of the 13 patients (77%), there was reported improvement in the symptoms and signs of an FOP flare-up. In three of the 13 patients (23%), neither the physician or the patient detected improvement in the symptoms of the flare-up. Interestingly, there seemed to be no protective effect whatsoever on the occurrence of subsequent flare-ups in any of the patients treated with either a single dose or a brief course of intravenous Pamidronate.

The treatment protocols varied slightly among the patients (depending on age, body weight, and site of involvement) but in general were similar. The most commonly used protocol, as well as additional discussion of this medication, is published in "The Medical Management of Fibrodysplasia Ossificans Progressiva: Current Treatment Considerations."

Generally, for treatment of acute flare-ups involving major joints, the FOP treatment guidelines recommend considering a 4-day course of oral prednisone in conjunction with a 3-day cycle of IV (intravenous) Pamidronate (often starting prednisone a day or two prior to the administration of Pamidronate), though circumstances vary by patient and flare-up). If swelling recurs following the discontinuation of prednisone, a second 4-day course of high dose prednisone may be given with a slow taper of the prednisone over the following 10 days.

Side effects of the intravenous Pamidronate infusions in FOP patients typically include flu-like symptoms of fever, chills, and muscle aches. These symptoms can often be lessened by pre-treatment with acetaminophen. One patient developed tetany (uncontrolled muscle contractions due to a low vitamin D level in the blood prior to treatment), and one patient developed peripheral phlebitis (inflammation of the vein) at the intravenous infusion site, which required inpatient intravenous antibiotic treatment. A recently published case report documents the development of osteopetrosis (condition in which bones become abnormally dense) in a child treated with 60 mg of IV (intravenous) Pamidronate every three weeks for two years. The child did not have FOP.

One important cautionary note about the bisphosphonates is necessary. Osteonecrosis of the jaw (ONJ) has been increasingly suspected to be a complication of bisphosphonate therapy, especially recurrent intravenous administration of the more potent aminobisphosphonates, such as Pamidronate and Zoledronate. ONJ, a rare dental condition, is diagnosed when an area of exposed jaw bone shows no sign of healing eight weeks after an invasive dental procedure, such as a tooth extraction. The gum that would normally cover the bone deteriorates, and the underlying jawbone is exposed. Some patients experience discomfort in the affected part of the mouth. Antibiotics have been effective for some patients, but generally there is no effective treatment. Clinicians and patients should be aware of this potential complication and a patient's dentist should be made aware of any history of using bisphosphonate medications. Should people taking bisphosphonates for FOP be concerned? Perhaps, but bisphosphonates are used to treat millions of people, and only an exceedingly small number of patients have developed ONJ. However, a small risk is present, and a few precautions are recommended: a dental exam, if possible, before Pamidronate treatment should be considered. Pamidronate should be avoided, if possible within 8 weeks of major dental surgery.

There are some clues that may help indicate why Pamidronate therapy might be effective for FOP, but we still need to put all of the pieces of the puzzle together to determine whether it is truly helpful as a treatment. As a consequence of its potent suppression of bone resorption, the aminobisphosphonates effectively inhibit the release of certain

substances in the body, including bone-forming proteins. But if aminobisphosphonates inhibit FOP flare-ups by decreasing the release of bone-producing proteins sequestered in the skeleton, one would expect a more pronounced effect on the prevention of subsequent flare-ups based on the fact that aminobisphosphonates can suppress these substances for months to years. Aminobisphosphonates also have an anti-angiogenic effect (decreased new blood vessel formation) and can lead to decreased production of lymphocytes, special white blood cells that lead to inflammation and carry bone-producing cells in people with FOP. This also makes them potentially desirable for treating FOP.

All of us in the FOP community know that anecdotal observations could be purely coincidental—that is, that the flare-ups might have receded spontaneously without treatment and that the Pamidronate might have had nothing to do whatsoever with the reported improvement, especially since oral glucocorticoids such as prednisone were used at the same time in many of these patients. Also, one cannot discount a potent placebo (wishful thinking) effect in any uncontrolled observation. Nevertheless, we also know that such observations of potential improvement in an FOP flare-up cannot be ignored, and we will continue to investigate Pamidronate in laboratory and clinical situations.

The immune system and why montelukast (Singular) might be helpful in treating FOP

The medication montelukast (Singular) is typically used in the treatment of asthma because it helps suppress special substances called leukotrienes. Leukotrienes are products of the immune system. They are chemicals released by mast cells, which are basically the atom bombs of the immune system, carrying an arsenal of items to help our bodies fight off injuries and infections. Unfortunately, sometimes the bombs get set off when we don't want them to and cause unwanted inflammation and swelling. Montelukast hopefully keeps this process from happening quite as often. You are probably now understanding why researchers are thinking that montelukast (Singular) might be helpful in treating FOP. In some cases, it has been combined with a non-steroidal anti-inflammatory medication or a Cox-2 inhibitor as a long-term treatment following a flare-up. Some individuals take it daily in the hope that it might have a beneficial/potential effect on future flare-ups. There have been no formal studies to assess the drug's effectiveness for FOP.

See “The Medical Management of FOP: Current Treatment Considerations” for more information, including recommended dosages.

Muscle relaxants

During times of flare-ups, people with FOP often experience pain and muscle contraction and sometimes experience muscle spasm. These spasms can affect mobility as muscles contract. For these reason, use of muscle relaxants such as cyclobenzaprine (Flexeril), metaxalone (Skelaxin), or lisoral (Baclofen) may be helpful. This is especially true for painful flare-ups involving the major muscle groups of the backs, arms and legs. The chronic use of muscle relaxants between flare-ups (to help deal with the muscle restrictions caused by FOP bone) has not been widely reported, but has been tried by some individuals. Dosing schedules are especially important with certain muscle relaxants (such as Baclofen) and need to be tapered carefully to avoid side effects.

FOP and clinical drug trials

The “gold standard” for testing a medication to see if it is truly effective is a test called a double-blind randomized placebo-controlled study. In such a study, some patients take medication, while others take a placebo. Neither the patients nor the doctor know what each participant is taking until the end of the study. This is the only truly objective and scientific way to evaluate a medication in humans.

There have been no such studies for any of the medications used to treat FOP flare-ups, partly due to the fact that FOP is such a rare disease, partly due to the unpredictable nature of FOP, as well as other obstacles. However, there have been several open studies of various medications. Open studies are studies in which all participants took the medication being tested during the study.

What to do in common flare-up situations

Some of the most common clinical situations facing families with FOP and possible treatment considerations are summarized on the chart at the end of this chapter. *This information reflects the experience and opinions of the University of Pennsylvania research team and the International Clinical Consortium on FOP on medications which have either been used in the treatment of FOP. This information is meant only as a guide. Again, although there are common physical features shared by every person who has FOP, there are differences among individuals that may alter the potential benefits or risks of any medication or treatment option. The final decision about treatment must be made between you and your doctor.*

For more specific recommendations, including dosage information, see “The Medical Management of FOP: Current Treatment Considerations.”

The future of FOP treatment

The best hope for treating FOP lies in future identification of better treatment strategies. There is no doubt that the recent discovery of the FOP gene is the most valuable piece of information in the FOP puzzle (read more about this in Chapter 15, “The FOP gene”), but it is only the “cornerstone” piece. Researchers still need to understand more about how ACVR1 (the gene that is involved in FOP) works—in everyone as well as people with FOP—before they can develop effective treatments for people with FOP.

To develop an effective treatment for FOP, the FOP gene will have to be disabled, blocked, neutralized, or bypassed. Those who work on FOP research have often said that FOP research is like trying to figure out the wiring of an atom bomb so that the bomb can be safely defused before it explodes. The FOP mutation, or the trigger of the atom bomb, is now known. The next step is to determine how to safely deactivate it. This will take time. Development of medications used to treat rare “orphan” diseases is very difficult. Many obstacles can be encountered including issues of safety, drug tolerance, side effects, drug delivery (how to administer a drug, for example pill, liquid, IV/intravenous, cream, gene therapy, etc.), and determining how well a medication targets the problem. A lot of research and testing must be done. That is the sobering news. But the great news is that we now have an extremely specific target for drug development that will immediately focus an enormous amount of medical and scientific attention on this gene and on FOP. Researchers are hard at work investigating new strategies to treat FOP.

Potential treatment considerations for flare-ups and injuries

Situation	Treatment considerations
Falls and head trauma	<ul style="list-style-type: none"> • Locked upper limbs may accentuate head and neck trauma from falls. Epidural hematomas are common in severe falls and a surgical emergency. All head and neck injuries must be evaluated immediately by a physician. • Parents may wish to consider use of protective headgear for children.
Severe soft tissue trauma threatening use of a limb (for example, following a fall but before a flare-up occurs).	<ul style="list-style-type: none"> • Apply ice intermittently, as tolerated, to injured area for 24 hours. • May consider a brief 3-day course of prednisone. If flare-up subsequently occurs, consider treating symptomatically as indicated below. Do not use prednisone after minor bumps and falls.
Flare-up involving back and/or chest	<ul style="list-style-type: none"> • May consider symptomatic treatment with a non-steroidal anti-inflammatory medication or Cox-2 inhibitor (celecoxib) with gastrointestinal precautions to prevent stomach issues. Use analgesics (pain relievers) and/or muscle relaxants as needed. • Prednisone should not generally be used for the treatment of flare-ups involving the back, neck, or trunk due to the long duration and recurring nature of these flare-ups, and the difficulty in assessing the true onset of such flare-ups. On rare occasions, a brief course of corticosteroids (prednisone) may be used to break the cycle of recurrent flare-ups often seen in early childhood. However, the usefulness of this approach is not widely accepted, as flare-ups tend to recur rapidly following stopping corticosteroid therapy.
Flare-up involving limbs	<ul style="list-style-type: none"> • May consider brief 4-day course of prednisone. Begin within first 24 hours of flare-up. Keep medication on hand for emergencies. Use analgesics (pain relievers) and/or muscle relaxants as needed. Take gastrointestinal precautions. • A 2-3-day course of IV (intravenous) Pamidronate infusions may be considered in conjunction with prednisone for acute flare-ups (often starting prednisone a day or two prior to the administration of Pamidronate). Zometa (zoledronic acid) may also be considered for those 18 years of age or older. Zoledronic acid should not be used for younger patients.

<p>Flare-up involving submandibular area (underneath jaw)</p>	<ul style="list-style-type: none">• Strict avoidance of manipulation or repeated palpation• Airway monitoring• Aspiration precautions (Aspiration is an audible breath that compromises speech.)• Nutritional support• May consider use of prednisone as above with a long taper (3-4 weeks or until flare-up subsides) to decrease soft tissue swelling to this vulnerable area if airway appears threatened, or if swallowing is impaired. This is one of the few situations in which a more prolonged use of corticosteroids is justified. Prednisone may also be used in conjunction with Pamidronate or Zoledronate.
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7. Emergencies

Follow these guidelines when dealing with emergency situations. Please also see Chapter 5, “The medical world,” for several ways to help educate medical professionals in the event of an emergency.

Evaluating an emergency

If an emergency medical situation develops, whether a bad fall, a broken bone, appendicitis, or something else, a good rule to follow is for your local doctor or emergency department personnel to evaluate the problem and then contact Dr. Frederick Kaplan or Dr. Robert Pignolo at the following:

Frederick Kaplan, MD
 (215) 349-8726/8727 (office)
 (215) 545-0758 (home)
 Frederick.Kaplan@uphs.upenn.edu

Robert Pignolo, MD
 (215) 349-8726/8727 (office)
 (215) 308-9643 (pager)
 Pignolo@mail.med.upenn.edu

Please Note: Dr. Pignolo is also available by “text paging,” which allows you to send him a text message through the online service USA Mobility. To “text page” Dr. Pignolo, visit www.usamobility.com, choose the “Send a Message” link, enter his pager number, write a short message and your contact information, and submit.

For a complete list of doctors around the world please visit Chapter 28, “Medical specialists around the world.”

Most emergencies that people with FOP have are not related to their FOP, but to common problems that anyone would have. Treatment should take the special needs of FOP into account.

The following simple rules apply:

- Intramuscular injections should be avoided as they may cause an FOP flare-up.
- Medications can be safely administered intravenously (into a vein) if needed.
- Special care should be taken to avoid unnecessary trauma, including medically unnecessary surgery.

If surgery is needed

While surgical operations usually make FOP worse by causing the body to generate more bone, emergency situations may exist where an operation may be necessary, as in appendicitis or acute gall bladder disease. Although such an operation may lead to a flare-up, the life-threatening nature of the emergency may justify the surgery.

The following guidelines may be helpful in dealing with surgery during emergency situations. (These are a modified version of the guidelines listed earlier in this book.)

- Be aware of the risks of new bone formation following injury to, or surgery on, the musculoskeletal system.
- Avoid injury to the musculoskeletal system.
- Again—avoid intramuscular injections, including local anesthesia.
- Careful planning and management of airway problems should be recognized and implemented.

General anesthesia, the type of anesthesia recommended for people with FOP, leads to an unconscious state in which a person is pain free and unaware of what is happening. It is often administered as an inhaled gas. General anesthesia is a particularly dangerous matter in people with FOP, so special precautions need to be taken. Overstretching of the jaw muscles for intubation (placing of a breathing tube into the trachea) may cause trauma to the muscles and joints of the jaw and lead to flare-ups. Airway complications can also occur if the body reacts to pain, mouth secretions, or bleeding by closing the vocal cords. This is a potentially life-threatening situation for anyone, and even more for someone with FOP.

Because of jaw fusion, typical intubation may physically not be possible. Another approach is called awake fiberoptic nasal intubation. With this procedure, an anesthesiologist guides a fiberoptic laryngoscope (a type of tiny medical camera that can look inside the body) through the nose in an effort to indirectly visualize the airway while a person is awake. (Complete sedation is not recommended for people with FOP because of potential anesthetic complications and pulmonary/breathing issues).

Once the airway is visualized and the patient is intubated, then general anesthesia can be administered. It is important to note that fiberoptic intubation should only be performed by trained anesthesiologists who are experienced with this type of procedure.

For questions related to anesthesia, emergency room personnel should contact Dr. Zvi Grunwald at 215-955-6161 or zvi.grunwald@jefferson.edu. Dr. Grunwald has significant experience in dealing with the anesthetic needs of people with FOP.

How to handle other injuries

If an injury occurs (such as a sprained muscle, pain from a fall, getting bumped, etc.) but does not seem serious enough to require an emergency room visit or a visit to the doctor's office, consider applying ice to the injury site as soon as possible. Ice helps minimize inflammation and swelling. Cold has the added benefit of being a good natural pain reliever. Follow this procedure to help the recovery process from an injury:

RICE, which stands for ...

R	Rest	Minimize movement of injured body part
I	Ice	Apply a cold pack
C	Compression	Apply light pressure to the affected body part
E	Elevate	Raise the body part to help decrease swelling

Ice has maximum benefit in the first 48 hours following an injury. If ice does not resolve the inflammation and pain, you may find yourself in a position where you need to make a judgment call regarding whether you are dealing with an injury alone or whether the injury has become an FOP flare-up. Unfortunately, this is not always an easy determination to make, and you will have to learn how to trust your best judgment. Generally if you think it is an injury, it is best to continue treating it as an injury. But if you believe it is a flare-up, then follow the recommendations in Chapter 6, "Making FOP treatment decisions."

If a soft tissue trauma occurs that threatens the use of a limb, you may also consider a brief 3-day course of prednisone. If flare-up subsequently occurs, consider treating symptomatically as indicated in Chapter 6, "Making FOP treatment decisions," and as described in more detail in "The Medical Management of FOP: Current Treatment Considerations." Do not use prednisone after minor bumps and falls.

8.

Treating acute and chronic pain

People with FOP often experience pain during flare-ups as new bone invades muscles and connective tissue. Some individuals, particularly those with more advanced cases of FOP, also experience chronic pain that does not go away. Not everyone experiences chronic pain. Learn the basics of treating pain so that you can work with medical professionals to find a way to control pain and make life more bearable.

Introduction

To sum it up simply, the goals of pain management are to lessen pain, improve a person's ability to function and do the things that he or she enjoys, and to enhance one's quality of life. Unfortunately, the process of controlling pain, particularly chronic pain, isn't always easy. That is why it is important to know about available treatment options and to work with medical professionals until you find something that works.

A little note about grammar (Don't worry. This is not a grammar lesson!): Many areas of this chapter make use of pronouns such as our or you (or your child) or our. Ours is often used for principles that relate to all of us, whether we have FOP or not. The term you or your child is used in reference to dealing with pain or working with medical professionals. Sometimes you is also used to note that pain affects not only one person, but the whole family, and the whole family may be involved in helping to find the best treatment options.

The information provided in this chapter is meant to complement and not replace any advice or information from a health professional.

Keep these things in mind

Keep these things in mind as you seek treatment for pain:

- Be sure to seek treatment as early as possible before pain becomes unbearable.

- Consider keeping a pain journal that can be shared with your child's (or your) physician. Continue reading this chapter to learn more about keeping a pain diary.
- Before going to doctor appointments, write down a list of questions that you want to ask and provide doctors with information about FOP (treatment guidelines, a personalized "Medical World Binder," guidebook, etc). This helps you make the best use of the time that your child's (or your) doctor can spend with you.
- Accept support from loved ones.
- Consider bringing a relative or friend to your appointments to provide support or to help you remember details.
- Make sure someone from your healthcare team is available if you need to call with questions about medications or other concerns.
- Know about available treatment options.
- Attitudes and expectations make a difference. Try to stay positive.
- Learning to relax is very important. Our bodies and minds are linked—emotions affect how we feel. Relaxing helps prevent muscle tension and redirects our thoughts onto the things within our own control.
- Stay as active as possible. This can help keep our thoughts off pain. It also helps us to feel more in control of our lives.
- Set realistic goals. We all learn to walk before we can run. Similarly, pain management is often a process that may take time.

Available options and where to turn for help

As the American Pain Foundation states, "Pain is complex and unique to each individual. For this reason, your healthcare team will consider many aspects of your pain and daily life before recommending a treatment." Some of the things to be considered include the type of pain, for example whether it is acute and based on a current injury or problem, whether it is chronic pain that doesn't go away, the intensity of pain, a person's physical condition, lifestyle, and treatment preferences.

Some pain problems can be managed by an individual's personal physician, so this is always a good place to start. When pain is more difficult to treat, a person may be referred to healthcare professionals who specialize in treating pain, for example neurologists, anesthesiologists, and some psychiatrists. A person might also be referred to a clinic that specializes in treating pain. Potential approaches for controlling pain might include medications, lifestyle changes, improved coping strategies, counseling, and complementary/integrative medicine.

To find a pain specialist, try the following options:

- Ask your doctor for a referral to a pain specialist.
- Ask family members or friends who have had pain for suggestions.
- Contact the largest hospital in your area.
- Call government health agencies and resources (country, state, county).
- Contact a hospice.
- Contact professional organizations for pain specialists. Two such organizations are the following:
 - American Academy of Pain Medicine
www.painmed.org
 847-375-4731
 - or
 - American Pain Society
www.ampainsoc.org
 847-375-4715
- Contact an organization that provides information about dealing with pain:
 - American Chronic Pain Association
www.theacpa.org
 800-533-3231
 - or
 - American Pain Foundation
www.painfoundation.org
 888-615-PAIN (7246).

Once you have found a specialist, contact his or her office to learn more about that person's approach to managing pain and to schedule an appointment if you feel that approach will help your child (or you). Before your appointment, read the additional parts of this chapter to learn more about potential treatment options so that you can have an informed conversation about how to best manage your child's (or your) pain.

Pain relationships

Pain can impact our emotions, and our emotions can impact pain. In other words, pain may make us feel stressed, sad, or anxious. And feeling anxious or sad can interfere with recovery or make pain worse. As an understanding of pain improves, doctors are realizing that influencing emotions can help with the treatment of pain.

Possible strategies that may be effective in controlling pain by affecting our emotions may include:

- Relaxation therapy – Practicing forms of meditation, which help a person to be more aware of his or her breathing; may include guided imagery (relaxation and visualization of mental images that are pleasing)
- Biofeedback training – Teaching people how they respond to stress through the use of special equipment that monitors brain activity, blood pressure, muscle tension, and heart rate. This training is then used to help them correct patterns that can lead to pain.
- Behavioral modification – Changing attitudes and habits that may be contributing to pain.
- Stress management – Learning to set a routine schedule. (Not knowing what to expect can cause stress.) Find an activity that you like and make it part of your daily routine. Talk positively and celebrate the good things in your life, even small accomplishments. Think about the people who enhance your life in good ways. Learn to live in the moment and try to set aside negative thoughts.
- Counseling (individual or family, as pain often affects family dynamics as well) – Pain can produce feelings of hopelessness, fear, and anger. Sometimes talking to a professional counselor can help. When choosing a counselor, it is good to find a person with experience in pain management.

Complementary medicine

Conventional western medicine is starting to pay more attention to combining standard medical treatment with other forms of treatment. This is called complementary or integrative medicine. The National Institutes of Health in the United States recently opened the National Center for Complementary and Alternative Medicine to help evaluate these therapies. Many of these forms of therapy are holistic, meaning that they look at mind, body, and spirit in treating a person.

To learn more, contact them at

National Center for Complementary and Alternative Medicine
888-644-6226
www.nccam.nih.gov/health

Please consult your physician before pursuing complementary and alternative medicine.

If interested in pursuing any of these therapies, the following suggestions may help you decide which is most appropriate for your needs and help you find a practitioner:

- Ask your doctor for recommendations. A nearby hospital or medical school may also maintain a list of practitioners or might be able to make a recommendation. Some medical centers may have integrative health centers or practitioners on staff.
- Contact a professional organization for the type of practitioner that you are seeking. Resources include the following:
 - International Association of Healthcare Practitioners
800-311-9204
www.iahp.com/pages/search/index.php
(lists practitioners who specialize in various therapies)
 - dirline.nlm.nih.gov
(which maintains information about various professional organizations)
 - www.amtamassage.org
877-905-2700
(which lists therapists affiliated with the American Massage Therapy Association and shows which types of massage they practice)

If you do a web search, be very general about your location. For example, entering a state or country will give you more names, but you may find someone near you that would not have appeared if you had tried a more specific search by city. If you have internet access, you may also learn more about a type of therapy and available practitioners in your area by doing an internet search.
- Many states have licensing boards for certain types of practitioners. Contact your state, county, or city health department for more information.

Once you have found the name of several practitioners, call each one and ask the questions found below. Also explain FOP as simply as possible so that they can understand your needs better. For example, you might say that FOP causes bone to grow in muscles and connective tissue and can significantly affect movement.

Here is the list of questions to ask practitioners and therapists:

- What type of training have they had? How many years have they practiced?
- What types of health conditions do they generally treat? Ask if they feel their therapy would help you and if they would be comfortable treating you, as treating someone with FOP can involve more creativity than treating the average person. (From your conversations, you should get an idea of how flexible each person is.)

- Is there a website you can visit for more information? Do they have a brochure?
- How long is a session and what is the cost? (In most cases, these therapies are not covered by insurance.)
- How often do they recommend sessions be scheduled? How far in advance will you typically need to make appointments?
- If needed, ask if the office location is wheelchair accessible. You may wish to ask if the person is willing to make house calls. Some will, some won't, and there is usually an extra cost for this.
- Ask what type of massage table is used. For example, is it height adjustable for easier access? If they are willing to do house calls, would they be comfortable using a bed, as many portable massage tables do not adjust to different heights.
- Ask what will happen at the first visit.

An important note: A person with FOP should instruct the therapist to be as gentle as he or she can while still being effective. After the first visit, it is also good to evaluate whether you felt comfortable working with the person and whether you felt the type of treatment might be helpful to you. In many cases, this is hard to know with certainty until you have had a therapy session. For instance, the therapy may be appropriate but the therapist is not, or vice versa.

Pain diary

A pain diary can be a helpful tool in managing pain. It will help identify when pain was bad and what made it seem better. Don't worry about how much to write. Don't worry if you miss a day or a few days because you don't feel up to writing anything—just come back to it when you are ready or perhaps ask a friend or relative to help you.

Write down the date and time of each entry. You may wish to use the following questions as a guide to how you are feeling:

- Where does it hurt?
- How does the pain feel?
- Did the pain exist when your child (or you) woke up, or did it start later?
- Does the pain change during the day?
- What makes the pain better or worse?
- What medications is your child (or you) taking? Do they help?
- Does your child (or you) have trouble sleeping?
- Does the pain interfere with activities?
- Does the pain interfere with eating?

The American Pain Foundation's website features a downloadable pain journal, as well as other resources that can be downloaded, at www.painfoundation.org. Visit their publications section for more information.

Conclusion

Understanding and managing pain is not easy. But do not give up until you have an effective pain management program. There are many places that you can turn for help. One of them is the American Pain Foundation. While the information in this section was compiled from various sources, a particularly helpful resource was the American Pain Foundation's publication *Treatment Options: A Guide for People Living with Pain*, which can be downloaded from the publications section of their website at www.painfoundation.org or obtained by calling 888-615-PAIN (7246). The document contains many additional resources for people in pain.

Other resources include the following books, which can be obtained through Amazon.com. Descriptions come from Amazon.com and publishers.

- Barksy, Arthur J. and Deans, Emily C. *Stop Being Your Symptoms and Start Being Yourself: The 6 Week Mind-Body Program to Ease Your Chronic Symptoms*. Dr. Arthur Barksy, psychiatrist and pioneer in the field of mind-body medicine, has found that changing the way you think about your illness can have a remarkable effect on how you experience your symptoms. At Harvard Medical School, Dr. Barksy developed Stop Being Your Symptoms and Start Being Yourself, a program designed to overcome the symptoms of chronic illnesses of every kind. This groundbreaking program teaches patients to master the five psychological factors that make chronic symptoms persist through hundreds of exercises, worksheets, and patient examples. You may not be able to completely eliminate your medical symptoms. But it is possible to control your symptoms rather than letting them control you—to manage your pain, fatigue, insomnia, and anxiety. You can minimize your symptoms, learn new coping skills, and do more to make sure that your symptoms are not robbing your life of meaning and pleasure.
- Claudill-Slosberg, Margaret and A., *Managing Pain Before It Manages You*. “Dr. Caudill’s narrative is crystal clear and insightful, demonstrating great empathy for pain sufferers. She has a knack for clearly and concisely discussing relevant information while anticipating readers’ misconceptions and reservations, addressing them throughout the volume. This volume can be used by people with chronic pain on their own as well as an adjunct to treatments provided by

- professionals. If followed carefully, this volume will surely make a difference in the lives of those with diverse chronic pain syndromes. Dr. Margaret Caudill has produced an outstanding volume that will set the standard against which all future volumes targeting those with chronic pain will be judged.”
- Cochran, Robert T, Jr. *Understanding Chronic Pain: A Doctor Talks to His Patients*. This is a personal narrative, a record of my passage among victims of chronic pain and the discoveries that have come from those encounters. I write for physicians, nurses, therapists, and caregivers, but mostly, I write for you who suffer the disease. I know you very well, perhaps as well as anybody in the world. I have listened to your stories with patience and attention, and I have been greatly rewarded. You have trusted me with the deep recesses of your thoughts and fears, and the memories of the dreadful experiences that are so often the origin of chronic pain. I have treated thousands of you and I believe I have some understanding of your illness. I offer a series of essays about people like you who suffer chronic pain. From their case histories, I derive certain conclusions. Some conclusions are bold and imaginative. Some are disturbing and frightful. Not all of them will apply to you, but some certainly will. My wish is that you gain greater understanding of your illness, for only understanding it will you conquer it.
 - Turk, Dennis and Frits, Winter. *The Pain Survival Guide: How to Reclaim Your Life*. If you suffer from chronic pain, this proven 10-step program brings hope and relief, showing you how gradual changes in specific behaviors can lead to great improvements in your ability to cope. The authors help you learn “not to let your body push you around” so life becomes enjoyable again. The key lessons in this book include • Uncovering some of the myths about pain and the deceptive ways it fools your body into unconstructive behavior • Pacing your activity, so you build strength without overdoing or underdoing it • Learning how to induce deep relaxation so you can begin to enjoy life again • Dealing with disturbed sleep and chronic fatigue • Improving your relations with family and friends, and soliciting support • Changing your habitual behaviors in ways that reduce pain • Combating the negative thinking that often accompanies pain • Regaining your self-confidence and trust in yourself • The power of goal-setting and humor • Dealing with the inevitable relapses and setbacks once improvement has set in Workbook exercises, behavior logs, and suggested readings help you integrate these lessons into your daily life and learn to live well despite pain.

There are many other books about pain relief. Which book may be most helpful will be a personal preference, but hopefully this information gives you some ideas.

9.

Oral and dental concerns

When the jaw becomes affected by FOP, the issues of eating and dental hygiene come to the forefront.

When does the jaw typically become affected?

The joint in the jaw, referred to as the temporomandibular joint (TMJ) is typically one of the later joints to be affected by FOP. However, FOP involvement of the jaw muscles has been reported following dental procedures or oral trauma (accidents, overstretching of the jaw, etc.) at any age. In a study of people with FOP, 71% reported jaw restrictions by age 18.

Preventive dental care

Preventive dental care, which involves making an effort to avoid cavities and keep the teeth and gums healthy, is extremely important for individuals with FOP. If the muscles and joints of the jaw are fused, then teeth cannot be brushed on all surfaces and individuals have a higher likelihood of developing dental problems such as cavities or gum disease. Good preventive care routines can help minimize the chances of such problems occurring.

Here are some guidelines to follow:

- All affected individuals should have early, regular, and periodic dental visits combined with oral hygiene instruction. Individuals who still have full mouth opening can be treated with regular dental instruments, though special care should be taken not to overstretch the jaw. For those with restricted jaws, dental specialists such as special needs dentists (who specialize in treating people with disabilities) or periodontists (who specialize in gum disease and are used to dealing with people with many severe dental problems) may have unique training or equipment that can better meet the needs of people with FOP.
- The use of dental sealants (applying a special material to the surfaces of molars and back teeth to help prevent tooth decay) might be considered before jaw fusion happens. Typically sealants are applied during childhood, but this work may also be done at any age as long as the teeth are still accessible to a dentist.
- Frequent brushing with high fluoride toothpaste is recommended along with the use of fluoride gels or rinses. Chlorhexidine rinses are also recommended.

Chlorhexidine is an anti-bacterial rinse that can help prevent gingivitis (gum inflammation) and tooth decay. Rinses can also help a person access areas which otherwise might be difficult to reach because of jaw fusion.

- Many individuals have found it helpful to use electric toothbrushes such as those made by Sonicare or Braun/Oral B. In addition, electric flossing systems allow individuals with jaw restriction to floss more easily. Waterpiks (electric water rinsers) may also be useful.
- A new product is MI Paste (made by GC America). It remineralizes enamel (the protective coating on teeth) to prevent tooth decay and gum disease, particularly in people who cannot brush their teeth adequately. Do not use this product if you are allergic to milk proteins.

Eating

When extra bone forms in the muscles and connective tissue of the jaw, it can understandably become difficult to get food into the mouth. Some people with fused jaws find that they can still eat food that is cut into small pieces, while others find they need to stick with very soft foods or puréed foods. Like most aspects of FOP, there can be great variability in how the muscles and joints of the jaw fuse.

Sometimes, an FOP flare-up in the jaw or chin area may also make it temporarily difficult to eat, or eating may aggravate the flare-up. In these cases, a person with FOP may need to take special high calorie dietary supplements such as Ensure or Boost so that body weight can be maintained. If Ensure or Boost are unavailable in your country, ask your doctor about available food supplements.

If there is a way to eat it...

If there is a way to eat something, a person with FOP will probably find it. After all, who wants to miss out on his or her favorite foods? The trick to eating with a fused jaw is learning to be creative. For example, in some cases it may be helpful to use a food processor, blender or food chopper to blend items to a suitable consistency.

Here are some tips for blending foods.

- Work slowly because blades work very quickly. If you “pulse” your food (stopping and starting the machine), you will have better control over the consistency of the food. Once you become more accustomed to blending foods, you will likely start learning how long specific foods take to blend to your desired consistency.

- Make a purée by beginning with enough liquid to cover the blades. Liquids can include milk, cream, broth, fruit juice, gravy, sauces, etc. Water may also be used, but it may dilute the taste. Gradually add solid ingredients.
- If you need to liquefy the food, maintain an equal amount of solids and liquids. If necessary, add additional liquid. You will need to add enough liquid to create a consistency that can be poured into a glass, sipped through a straw, etc.
- Blend foods before heating. If you don't, you might have to heat the meal again after blending.
- If using prepared or frozen food, cook it as you normally would. Then put the item in a blender, add a cup and a half of milk or broth. To perk up the flavor, a little salt and pepper, butter or sour cream. Add more liquids as necessary. You may want to put the final result through a fine mesh strainer. Experiment. Remember that this will be a learning process!

If you are looking for recipes that feature soft foods that are easy to chew, try the following cookbooks. Descriptions are from the publishers and/or the Amazon.com website.

The Dysphagia Cookbook: Great Tasting and Nutritious Recipes for People With Swallowing Difficulties. Elayne Achilles.

- *The Dysphagia Cookbook* is a specialty cookbook filled with nutritious, great-tasting recipes for those whose eating options are limited by chewing and swallowing difficulties. All of the recipes focus on enhancing flavor, presentation, texture, aroma, and color, for there are many other products that supply nutritious calories or liquids but give little attention to these quality-of-life concerns.
- Some of the unique aspects of *The Dysphagia Cookbook* make it particularly useful and practical. These include:
- A classification of S, G, or P indicates consistency levels of soft, ground, or puréed.
- Flexible instructions provide suggestions for adapting recipes to accommodate increasing levels of chewing and swallowing difficulties.
- There are many recipes with an international flavor that do not use difficult-to-find ingredients.
- Ready-made products that have been tested for thickness, flavor, ease of chewing, and ease of purchase and preparation are listed.
- A section on must-have kitchen supplies helps cooks deal with the new ways of preparing food for people with swallowing difficulties.
- The approach is practical rather than clinical.

- The ritual of eating gives shape and meaning to our lives. Many meals are consumed in a pleasant atmosphere with the company of loved ones and friends in lively conversation. *The Dysphagia Cookbook* is an attempt to restore this joy and dignity to those whose pleasure in this area has been limited to one degree or another.

Easy-to-Swallow, Easy-to-Chew Cookbook: Over 150 Tasty and Nutritious Recipes for People Who Have Difficulty Swallowing. Donna L. Weifhofen, JoAnne Robbins, Paula A. Sullivan

- The simple act of eating is a challenge for millions of people whose ability to chew and swallow has been compromised by the debilitating effects of age or disease. *The Easy-to-Swallow, Easy-to-Chew Cookbook* presents a collection of more than 150 nutritious recipes that make eating enjoyable and satisfying for anyone who has difficulty chewing or swallowing. It also shares helpful tips and techniques to make eating easier for the elderly and those with such diseases as Parkinson's, AIDS, or head and neck cancers.

The I-Can't-Chew Cookbook: Delicious Soft Diet Recipes for People with Chewing, Swallowing, and Dry Mouth Disorders. J. Randy Wilson.

- This book by J. Randy Wilson is a unique, one-of-a-kind cookbook that is an invaluable addition to the kitchen cookbook collection of any family chef who must prepare meals for anyone suffering from problems arising from a chewing disorder such as temporomandibular joint (TMJ) problems, stroke, ALS, Alzheimer's, AIDS, lupus, recovering from head or neck surgery, or mouth/throat cancer surgery. Randy Wilson drew upon his love of cooking and his array of culinary skills when his wife was diagnosed with TMJ and needed surgery. Her doctor indicated that she would have to subsist on soft foods for six months. Randy took on the challenge of developing soft, nutritious, and appealing recipes for his wife and their family. The result is his *I-Can't-Chew Cookbook*, which is neither a liquid diet book nor a blender cookbook, but instead showcases 200 soft and tasty recipes for casseroles, soups, entrees, side dishes, beverages, and deserts. Of special interest are the opening chapters on nutrition and tips for getting the most out of meals, including enhancing the dining experience and adapting foods for a soft-food diet when dealing with problems of swallowing and/or chewing. Enhanced with an informative Foreword by oral surgeon Mark A. Piper, the *I-Can't-Chew Cookbook* should be considered as a "must" for anyone wanting nutritious, delicious, consumable dishes for the chewing and/or swallowing impaired.

So What If You Can't Chew, Eat Hearty!: Recipes and a Guide for the Healthy and Happy Eating of Soft and Pureed Foods. Phyllis Z. Goldberg.

- This volume offers the keys to good eating when a soft or pureed diet is indicated. Those who find chewing difficult due to oral infection, jaw trauma or reconstruction, effects of oral cancer treatment or extensive dental care will find indispensable information on the preparation of appealing, flavorful and nutritious meals. Introductory chapters explain the optimal use of blenders and methods for coping with various oral problems. The author then presents over 100 detailed recipes for meat/fish/poultry, soups, fruits and vegetables, beverages, eggs/cheese/yogurt, desserts, and complete meals. Potage St. Germain, Chicken and Mushrooms, and Apricot Whip are representative of the recipes presented. The dishes described have all been kitchen-tested for tastiness and ease of preparation. The caloric and protein value of each recipe is included. Suggestions for daily menu planning also are presented.

Whipping a Whopper. Nancy Sando.

- This booklet, written by a woman who has FOP, offers tips and recipes for preparing food that can be eaten by a person with a fused jaw. Available through the International FOP Association (IFOPA). Visit ifopa.org or call 407-365-4194 for more information.

Creating extra space

In very extreme cases of jaw fusion, a person with FOP may wish to consult a dentist or a prosthodontist about whether a procedure called enameloplasty (or recontouring) would be beneficial. Enameloplasty is a painless procedure in which a very small portion of the enamel is removed. Normally it is a cosmetic procedure, but in someone with FOP enameloplasty can be used in order to create a slightly larger opening of the jaw. Dentists or prosthodontists can examine a person to determine which portion of enamel can safely be removed to maximize the jaw opening and minimize cosmetic implications.

Before the procedure, an X-ray should be taken to determine the size and location of each tooth's pulp (the center of the tooth, which contains nerves and blood vessels). If the enamel is too thin or if the pulp is too close to a tooth's surface, it is not wise to proceed. The main risk of enameloplasty is that too much enamel is taken off and teeth become sensitive to heat or cold.

It is a good idea to perform the procedure in stages. This way, appointments can be kept short to avoid any strain on the jaw. It also allows for very careful assessment of the situation to prevent potential problems.

For people with weight loss, causing health concerns, removal of selective teeth may be a possibility. This is a major procedure requiring a skilled dental and anesthetic team.

Minimizing risk during dental procedures— getting a good start

If a dental procedure needs to be performed on someone with FOP, a good first step is to assemble a team of experts. This means finding a dentist or oral surgeon who is willing to listen to you and understand the unique challenges of FOP and, particularly if the area is difficult to access, ideally one who has performed procedures on other individuals with jaw fusion. Your dentist should be able to make a referral to the most appropriate person.

Performing the procedure in a hospital is strongly considered. Ideally, the best location is a major medical center that routinely handles trauma cases, which can involve complications similar to some of the issues of people with FOP (jaw restriction, potential anesthesia/airway problems, etc.). If complications happen, these facilities will be more capable of handling them than if the procedure were performed in an office setting.

Minimizing risk during dental procedures— anesthesia

During dental work, a patient receives anesthesia so that a procedure can be done without causing pain. For most individuals who do not have to consider FOP issues, local anesthesia, which applies anesthesia directly to the problem area, is often used. *However, there are certain circumstances where local anesthesia should not be considered an option for people with FOP.*

First, let's talk about when local anesthesia is okay. For upper and lower anterior teeth (located in the front of the mouth), a dentist can use infiltration, an injection of local anesthetic into the soft tissue next to the tooth being treated. Upper molars can also be numbed by infiltration or nerve block local anesthesia, though local infiltration is the safest technique for people with FOP.

However, local anesthesia is typically provided as an *intramuscular* injection for procedures involving mandibular posterior teeth (lower molars). *As there is clear association between intramuscular injection of local anesthetic during dental procedures and FOP flare-ups in the jaw, intramuscular injections should be avoided.*

So what option is available for lower molars? The alternative is general anesthesia. (If the next paragraphs sound familiar, it's because parts of this section are also included in "FOP and Emergencies." It's important enough to repeat here.) General anesthesia affects a person's whole body, not just numbing the area being worked on like local anesthesia. It leads to an unconscious state in which a person is pain free and unaware of what is happening. General anesthesia is often administered as an inhaled gas. When you think of major surgery, you probably think of general anesthesia.

General anesthesia is a particularly dangerous matter in people with FOP, so special precautions need to be taken. Overstretching of the jaw muscles for intubation (placing of a breathing tube into the trachea) may cause trauma to the muscles and joints of the jaw and lead to flare-ups. Airway complications can also occur if the body reacts to pain, mouth secretions, or bleeding by closing the vocal cords. This is a potentially life-threatening situation for anyone, and even more for someone with FOP.

A recommended approach (and a necessary one if a person's jaw is already fused) is an awake fiberoptic nasal intubation. With this procedure, an anesthesiologist guides a fiberoptic laryngoscope (a type of tiny medical camera that can look inside the body) going through the nose in an effort to indirectly visualize the airway while a person is awake or sedated so that a person is still able to control mouth secretions. (Complete sedation is not recommended for people with FOP on account of potential anesthetic complications and pulmonary/breathing issues.) Once the airway is visualized and the patient is intubated, then general anesthesia can be administered. It is important to note that fiberoptic nasal intubation should only be performed by trained anesthesiologists who are experienced with this type of procedure.

Minimizing risk during dental procedures— fixing the problem

When jaw opening is limited, it may be technically difficult to access the problem area for the needed dental care. If a filling is required, access from the front (or buccal) surface of the tooth may be necessary, and tooth decay can be removed with the use of a slow dental drill. Use of fluoride-releasing filling material is also recommended. The filling materials will bond to the tooth and release fluoride, preventing further decay.

In patients with fused jaws, dental extractions pose many challenges. For example, the tooth may only be accessible from the front side. It may also be necessary to divide the decayed tooth into pieces prior to removal. A dental instrument must also be placed inside the mouth to prevent tooth debris from falling into the mouth.

Orthodontics and FOP

Like many people, individuals with FOP may have problems that require orthodontics (braces). This can safely be done for people with FOP who have normal or nearly normal jaw opening. However, if orthodontic care is considered, it is recommended that brief appointment times be used to lessen the stress on the jaw muscles. All bonded appliances are recommended to avoid extra pressure on the jaws. Non-extraction orthodontics, which involves straightening a person's teeth without removing any permanent teeth, is also recommended. To prevent tooth extractions, it may be necessary to align the front (anterior) teeth while leaving any crowding of the back (posterior) teeth untreated.

People with FOP, even when the jaws are not fused, commonly develop an overbite for reasons that are poorly understood. In these cases, please strongly consider leaving the teeth alone. Many individuals with FOP find that, once the jaws fuse, the overbite allows a means of access for eating and oral hygiene.

Where to turn for advice

For advice about dental care and anesthesia concerns, contact the following individuals, who have experience in treating a significant number of people with FOP:

Burton Nussbaum, D.D.S., M SND RCs Ed
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and
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E-mail: zvi.grunwald@jefferson.edu

A final note

After reading about the potential dangers and unique challenges that FOP poses for dental care, you may be tempted to not seek care out of fear that the jaws may fuse or that a procedure simply can't be done. Not seeking care can be just as dangerous. It can lead to additional problems in adjacent teeth or even life-threatening infections.

As you've learned by reading everything else in this book, no two situations are the same and there are many sides to a story. In any situation, the best thing to do is to absorb as much information as you can so that you can feel that you are making your decision as wisely as possible. Seek out all the help you can from experts and heed their advice. Doing nothing is bad news!

10.

FOP and breathing

Because of chest restrictions caused by FOP, individuals may develop breathing issues which require special medical attention. Learn ways to encourage good breathing and how to deal with problems if they occur.

An overview

In addition to the toe malformations that are typically noted at birth, individuals with FOP also seem to be born with a congenital malformation of the joints that connect the ribs to the vertebrae in the spine. This causes some degree of chest restriction even before the appearance of extra bone, though these restrictions may not lead to any noticeable problems. Because of these restrictions, people with FOP are more likely to rely significantly on the diaphragm muscle for breathing. The diaphragm is the dome-shaped muscle at the base of the lungs.

As FOP restrictions increase, extra bone and body position may start crowding the muscles around the lungs and/or heart to varying degrees, limiting chest expansion and creating additional restrictions which interfere with breathing. Flow rate (pattern of breathing in and out, inhaling oxygen, exhaling carbon dioxide) may be normal even when volume (the ability to take deep breaths and let them out) is greatly reduced. In some cases (usually more advanced ones), the body may develop higher than normal levels of carbon dioxide. Symptoms can be mild or severe depending on the degree to which FOP bone affects the chest area. The more advanced a case is, the more likely breathing difficulties may become serious enough to require treatment and the more likely there may be some right side involvement of the heart.

A study

In order to understand this issue better, a study of 25 individuals with FOP ranging in age from 5 to 55 years was conducted at an international meeting of people with FOP. Only one participant was under 13 years of age. The study involved physical examination, tests of pulmonary (lung) function, as well as electrocardiogram and echocardiogram (heart-related) studies. Although severe limitation of the chest wall was found, physical examination of the heart and lungs appeared normal. There was no evidence of heart failure in anyone who was part of this study. Ten of the patients had slight evidence of electrocardiographic

abnormalities indicating increased stress on the right side of the heart (the part of the heart chamber that pumps blood to the lungs). These individuals were generally older and had significantly longer duration of FOP symptoms than those who had normal electrocardiograms. All individuals had severely limited chest expansion and lung capacity (how much air the lungs can hold) was significantly reduced from normal levels. However, the flow of air through the lungs was relatively normal. Despite the limited capacity of the lungs to carry oxygen, the blood was well saturated with oxygen, just as you would see in people who do not have FOP.

When to consult a lung specialist

It is a good idea for individuals who have FOP to be evaluated by a pulmonologist, a doctor who specializes in lung disorders, and take a few tests in advance of serious problems in order to develop a baseline for later comparison. These tests determine any breathing restrictions that exist by doing a few tasks such as having the patient breathe into a spirometer (which can measure the flow and volume of breathing), checking the level of oxygen in the blood, or perhaps using a blood test to measure carbon dioxide levels.

Depending on the level of problems, the following approaches might be recommended:

- Breathing exercises using an incentive spirometer. An incentive spirometer measures how well a person is filling the lungs with each deep breath. Deep breathing helps to expand the tiny air sacs deep in the lungs. It helps mobilize secretions (such as those that are expelled when you cough) and keep the lungs full, open and as mobile as possible. While a person can practice deep breathing without the use of a spirometer, the spirometer offers the advantage of measuring a person's breath. It can determine whether the situation is remaining stable, improving, or getting worse.
- Positive pressure breathing. Positive pressure breathing is a type of breathing which is assisted by a machine. It encourages a person to take a deep breath by pushing in air with each breath. It is typically used to increase the amount of air a person breathes, help loosen secretions and help a person cough better. The machine can also be used to help administer medications into the lungs. This treatment must be given by a licensed respiratory therapist.
- Chest physiotherapy. Chest physiotherapy offers a way to clear the lungs of excess mucus and secretions. Under normal circumstances, the lungs are kept moist with a thin film of fluid. During a chest infection, this fluid can increase and become thick. In normal situations, this excess is removed by coughing, but that may not be possible when the breathing muscles are restricted and/or weak. Chest

physiotherapy uses gravity and physical therapy to help move secretions out of the lungs and stimulate coughing. Chest physiotherapy may be performed by a respiratory therapist or physiotherapist. Parents may also be taught to administer chest physiotherapy.

- Monitored oxygen use. Used appropriately, supplemental oxygen can be very helpful in advanced cases. Oxygen use in individuals with FOP must be monitored, as unmonitored use is associated with a high risk of sudden death because sudden correction of oxygen in the presence of high carbon dioxide levels (which are sometimes present in people with advanced pulmonary disease) suppresses respiration (the flow of breathing).

Pneumonia

Pneumonia is one of the most common causes of death in individuals who have FOP. It is part of the reason for the relatively low median life span of 41 years. (The term median simply means that half of people with FOP die below this age, and half live to be older than this. Individuals with FOP who do not develop life-threatening complications can live to be in their 50s, 60s, and even 70s.)

So what is pneumonia and why is it so dangerous to people with FOP? Pneumococcal disease is an infection that can attack different parts of the body. It can infect the lungs, where it causes pneumonia. It can also invade the bloodstream. If it reaches the brain, it can cause meningitis. These are all very serious infections. According to the Centers for Disease Control, pneumococcal disease kills more people in the United States than all other vaccine-preventable diseases combined. One out of 20 people who get pneumonia dies from it, and the statistics are even worse when the bacteria invade the blood stream or cause meningitis. People with health problems such as FOP are more susceptible to pneumonia and may have more difficulty fighting the infection.

Fortunately, there is a vaccine that protects against 23 types of pneumococcal bacteria. (There is no treatment for viral pneumonia, which usually heals on its own.) The pneumonia vaccine typically needs to be given only once, though a person should consult with his or her doctor for specific advice. In some high-risk groups, revaccination after a number of years is sometimes recommended.

The pneumonia vaccine is very safe. About half of those who get the vaccine have mild side effects such as redness or pain where the shot was administered. Less than 1% develop a fever, muscle aches, or a more severe local reaction at the injection site. Normally the vaccine is given intramuscularly, so the main precaution for individuals with FOP is that the vaccine

be administered as a subcutaneous (under the skin) injection. Make sure your doctor is aware of this special need. If any side effects result, contact your doctor as soon as possible. Side effects can often be managed with ice or anti-inflammatory medications but may require additional treatment.

Encouraging good breathing

Certain activities can help a person make better use of the muscles around the lungs and keep the chest muscles active. This will generally allow a person to get more oxygen with each breath and hopefully will lead to better breathing with less effort. In addition to practicing breathing with an incentive spirometer (see above), other activities that can help include deep breathing/relaxation exercises, swimming/hydrotherapy, singing, playing a wind instrument (trumpet, flute, harmonica, etc.) or even a toy instrument (like a kazoo, a toy that makes funny music when you breathe into it). Young children can try simple activities like blowing bubbles using deep breaths, or toys such as the ones listed in the IFOPA's *Catalog of FOP Resources*, available at the IFOPA website at www.ifopa.org. Laughing can also be helpful and is something that everyone can do. Laughing exercises the muscles of the diaphragm, abdomen and lungs as well as facial, leg, and back muscles. In some respects, it is like getting an aerobic workout. Laughing leads to deeper breathing that sends oxygen-enriched blood and nutrients throughout the body.

11.

FOP and the flu

A study conducted by FOP researchers has shown an intriguing relationship between influenza and FOP flare-ups. This recently established but still poorly understood link makes it especially important for people with FOP to avoid the flu. Learn more about this association, as well as ways to avoid getting ill.

Influenza

Influenza, commonly referred to as the flu, is a contagious respiratory virus. The symptoms can range from mild to severe and include the following: fever (usually high), headache, tiredness (can be extreme), dry cough, sore throat, runny or stuffy nose, muscle aches, and sometimes nausea, vomiting or diarrhea (stomach symptoms are more common in children than adults). Having these symptoms does not always mean you have the flu. Many different illnesses, including colds, have similar symptoms.

The flu is generally spread when people who are infected cough or sneeze. Occasionally people will become infected by touching something that has a flu virus on it and then touching their mouth or nose. Unfortunately, a person may be able to infect others beginning one day before symptoms appear and up to five days after becoming sick.

Increased risks for people with FOP who get the flu

People with FOP are at a higher risk for developing complications from the flu. The most serious complication is the development of respiratory infections such as pneumonia, which can require hospitalization and be potentially life-threatening. This type of complication happens as a result of the chest and breathing restrictions that are caused by FOP. A recent study has also shown that the flu serves as a potent trigger for FOP flare-ups. In fact, 60% of individuals in the study who had influenza also had flare-ups during the course of their illness, while only 11% of individuals who did not have the flu experienced flare-ups during the flu season.

It is not yet known why the flu seems to trigger FOP flare-ups. But since there does seem to be a link, it is possible that at least one trigger of FOP flare-ups may be based in the

immunological system. This explanation would make sense, as swelling and inflammation are immune system reactions. But at the present time the immunological features of FOP are still poorly understood.

Prevention is the best medicine

The best way to protect yourself from the flu is to get vaccinated each year. The flu shot contains an inactivated vaccine that offers protection from the types and strains of viruses that scientists determine will be circulating that flu season. The shot is given with a needle, usually in the arm. *It is especially important for a person with FOP to ask that it be given subcutaneously (under skin) with a very tiny gauge needle rather than into a muscle. (Remember that injections into a muscle are dangerous for people who have FOP!) In addition, an ice pack should be placed on the injection site and used intermittently for the next 12-24 hours in order to decrease the inflammation that can occur as a result of the injection.* Side effects from the flu shot may include soreness, redness or swelling where the shot was given, a low grade fever or aching. If these problems occur, they typically begin soon after the shot and last for one to two days. If you are allergic to chicken eggs (which are used to make the vaccine) or have had a severe allergic reaction to the flu vaccine in the past, you should not be vaccinated. *You should also not be vaccinated during an active flare-up.*

For those in the Northern hemisphere (United States, Canada, Europe, etc.), October or November is the best time to get vaccinated, but getting vaccinated later can still be helpful. Flu season can begin as early as October and may last as late as May. If you live in the Southern hemisphere (Latin America, Australia, etc.), the seasons are reversed, so flu season runs from May to September. So May or June is the best time to get vaccinated. For those of you who didn't know this, you didn't realize you were also going to get a lesson in geography, did you?

About two weeks after vaccination, your body will have developed proteins/substances called antibodies which will protect you from the flu.

Particularly if you dislike shots, you may be tempted to consider the new nasal flu vaccine. However, the nasal vaccine uses a "live" flu virus (rather than the inactivated virus in the shot), which *potentially* poses a higher risk to people with FOP. One young child who received the nasal flu vaccine developed a severe FOP flare-up two days after receiving the vaccine. This might have been coincidental or might have been due to the vaccine. It is impossible to say. Several other children received the nasal vaccine with no adverse effects. You should discuss all of the possible options with your doctor and make what you feel is the best decision for you.

In addition to getting vaccinated, the following precautions from the Center for Disease Control offer protection from the flu and other illnesses:

- Ask your family members and anyone else who is around you regularly to get vaccinated.
- Wash your hands often with soap and water. If you are not near water, you can use an alcohol-based hand sanitizer/antibacterial gel.
- Try not to touch your eyes, nose or mouth. Germs often spread this way.
- Cover your nose and mouth with a tissue when you cough or sneeze.
- If you get the flu, stay home from work, school or social gatherings so that others won't catch your illness.
- Avoid close contact from people who are sick. (And when you are sick, try to keep your distance from others.)

If you get sick

Since many illnesses have similar symptoms, it can be difficult to tell the difference between the flu and other infections. If you develop flu-like symptoms, contact your physician as soon as possible. Particularly because of the high risk of FOP flu complications, your doctor may recommend the use of one of several available antiviral medications (amantadine, rimantadine, oseltamivir, and zanamivir). These are medications which must be prescribed by a doctor and started within two days of illness. They will help minimize symptoms and speed recovery. Also get plenty of rest and drink plenty of liquids.

If someone else in the family gets sick, use of antiviral medications by other family members may help prevent spreading of the flu virus. In this way, antiviral medications can work on a preventative basis. Four studies that looked at two different antiviral medications showed them to be 75-80% effective in preventing someone who had been exposed to influenza from actually getting the disease.

12.

Kidney stones

Individuals with FOP may have a higher risk of developing kidney stones than those who do not have FOP. Learn what can be done to minimize this risk or handle this situation if it occurs.

What is a kidney stone?

A kidney stone is a hard mass developed from crystals that build up on the surface of the kidney. Normally our urine contains chemicals that help prevent this from happening. But sometimes problems occur. The most common type of stone contains calcium in combination with oxalate or phosphate. These chemicals are part of a normal person's diet and help form important parts of the body such as bones and muscles. In many cases, kidney stones are small and can pass through the body on their own. But when they don't, severe pain can result and treatment is necessary.

Kidney stones are one of the most common disorders of the urinary tract. According to the National Institutes of Health, approximately 5.2% of people in the United States developed kidney stones during the period from the late 1980s to the early 1990s, the most current period for which statistics are available. Kidney stones affect both sexes, but males are more likely to have diagnosed kidney stone problems. For men, the prevalence of kidney stones rises dramatically as they enter their 40s and continues to rise. For women, occurrence of kidney stones seems to peak in the 50s. If a person develops more than one stone, then others are likely to develop. Symptoms of kidney stones include abdominal pain, flank pain, pelvic pain localized to just one side of the body, a burning sensation during urination and/or blood in urine.

People with FOP and kidney stones

At the University of Pennsylvania School of Medicine, doctors started noticing that patients with FOP were asking about kidney stones on a more frequent basis than one might expect based on the estimated prevalence of kidney stones in the general population. Data from a study of 207 people from 31 countries, representing nearly half of known cases of FOP at the time the study was undertaken, were evaluated to determine if people with FOP have a heightened risk for development of kidney stones. Yet it is difficult to determine the specific risk of kidney stones in the FOP population with total accuracy because of geographic variation and the fact that international statistics for the prevalence of kidney stones are not available for all countries. (Geographic variation means that the prevalence of kidney stones

among people with FOP varied, sometimes significantly, in different countries.) Results did show that people with FOP are approximately twice as likely to get kidney stones than the general population in the United States.

The immobility caused by FOP combined with an increased bone turnover rate (the rate at which our bodies produce new bone and resorb existing bone so that our skeletons can remodel; something that may be affected by chronic decreased mobility) may play a role in these results. Urinary tract infections are also linked to kidney stone formation, in all individuals as well as people with FOP. Family history also influenced whether individuals developed kidney stones, particularly in males.

Diagnosis and treatment

Diagnosis of kidney stones is obtained by x-rays or sonograms/ultrasound, usually after someone complains of blood in the urine or sudden pain. Blood and urine tests may help determine which substances are present in the stone(s). The doctor may also decide to perform a special test called a CT scan (CT stands for computed tomography) or an IVP (intravenous pyelogram). The results of these tests will help determine the best treatment.

Sometimes no special treatment is necessary and a stone will pass through the body if large amounts of water are consumed. A doctor may prescribe pain medication to help a person feel more comfortable. A urologist will determine if treatment is needed. All available treatments, with the exception of percutaneous nephrostomy, have shown positive results for people with FOP.

Whenever anesthesia and/or surgery is required, remember to follow the guidelines listed in this book. See Chapter 2, “Things to avoid and alternatives,” on Chapter 7, “Emergencies,” for more information.

Recommendations

The most simple lifestyle change that a person can make to help prevent kidney stones is to drink plenty of water. Ideally you should drink enough water to produce 2.5 liters or more of urine each day. Try to drink as much water as you can, preferably around 3 liters each day.

Prevention is especially important for those who have had previous kidney stones, as there is a good likelihood that stones will form again. The following actions may also be recommended, especially for those who have had stones before:

- You don't need to restrict dairy, but don't overindulge either. People with kidney stones were previously told to avoid dairy products, but recent studies have shown that foods high in calcium may help prevent kidney stones. (However, studies have also shown that taking calcium in pill form may increase the risk of developing stones.)
- Avoid antacids which have a calcium base.
- If you have very acidic urine, you may be told to avoid food with added vitamin D. You may be asked to eat less meat, fish or poultry because these foods can increase the amount of acid in urine.
- If your urine has a high concentration of oxalate and you are prone to forming calcium oxalate stones, your doctor may ask you to limit the following foods in your diet: beets, chocolate, coffee, cola, nuts, spinach, strawberries, tea and wheat bran. Do not give up or avoid eating these foods without talking to your doctor.
- Do not consume high levels of vitamin C.
- Use whole wheat bread and natural fiber cereals.
- Limit salt in the diet.
- Use potassium magnesium citrate or other medications if needed. (Your doctor will recommend this if it is appropriate.)

Please discuss the topic with your doctor and decide if you need to take any special precautions to prevent the development of kidney stones.

13.

Miscellaneous health topics from head to toe

FOP involves many areas of the body in ways that go beyond the extra bone growth created by flare-ups.

Hearing loss

Loss of hearing occurs in a significant portion of people who have FOP. Based on patient surveys and medical records, it is estimated that 50% of individuals with FOP may be affected by some degree of hearing loss. As with many of the symptoms of FOP, there is variation. Some have more significant loss that requires the use of hearing aids, while others have minimal hearing loss. In some people, both ears are affected, while in others only one ear may be involved. In most cases, the hearing loss is conductive in nature, meaning that sound is not conducted properly from the ear canal to the eardrum and the tiny bones that make up the middle ear. In some cases the hearing loss is related to the nerves in the ear. There is much that is not understood about why hearing loss occurs in so many people who have FOP.

Although there is no direct association between hearing loss in people with FOP and childhood ear infections, it is important to remember that all children have a high likelihood of ear infections that can affect hearing. Therefore, it is important that earaches and other hearing problems in children with FOP be promptly evaluated and treated, as hearing loss from ear infections is a preventable problem in all children. Routine hearing tests are also recommended for people with FOP so that potential problems can be identified (even if you think your child hears well).

Ear and body piercings and tattoos

Body piercings and tattoos are very personal choices. Since these subjects are occasionally brought up on the FOPonline E-mail newsgroup, the topics are being addressed in this book even though they are not directly related to FOP.

Let's first address piercings. Piercings generally don't cause problems for people with FOP. However, it is a good idea to avoid the tongue, nose, and belly button. The tongue especially should be avoided because the tongue contains muscle and also because of the high likelihood of jaw fusion in people with FOP, which would make hygiene and/or removal difficult. Choose a reputable place if you wish to get a piercing and follow proper hygienic procedures to avoid infection.

Tattoos are the introduction of special ink into the skin using needles. Since a tattoo does not cause harm to muscles, it should not worsen FOP. However, the decision to get a tattoo should be considered very carefully because of the permanence of a tattoo. Anyone considering a tattoo should also choose a reputable facility that follows proper hygienic procedures. Improper administration of a tattoo can damage both your health and your skin. Allergy to ink can also be a problem for some people.

Headaches

Some individuals with FOP report chronic and severe headache symptoms. From a neuromuscular standpoint, this would make sense, as FOP causes significant restriction of the neck muscles which can contribute to tension-type headaches. If these headaches occur, there are medications that can help. A neurologist is the type of doctor who most often deals with chronic headaches that can be difficult to treat, and a neurologist will be able to make specific recommendations about the best treatment for a particular individual.

As with many issues relating to FOP, there is great variability, and many people do not report headache symptoms. Individuals who spend a lot of time at a computer should be careful not to stay in the same position for too long. Stop working from time to time to avoid muscle strain.

Milk is a good thing

Sometimes people will wonder if dairy products should be eliminated from the diet of a person with FOP because of the link between calcium and bone growth. Calcium is an important element for every person's body, even if a person has FOP. While it is well known that the calcium in milk helps build strong bones, it is not calcium that makes FOP bones grow. FOP bones grow because of a genetic signal in the body. Milk will not make the FOP worse, and it will keep the rest of the body healthy. Think about it this way. Both the regular skeleton and the extra one need to be fed with calcium. If a person with FOP does not ingest calcium, the bones will become weak and can easily break. While the body needs calcium for the bones to grow and stay healthy, it is also vital for other body functions. Calcium allows the nerves to function properly, helps the heart to beat, and contributes to other important

metabolic functions. The body cannot live without calcium. It is also especially important for people who have FOP to fulfill a recommended daily allowance for calcium because a strong skeleton offers better protection from injury if a person falls.

The importance of vitamin D

Like calcium, vitamin D is extremely important for keeping bones strong, and it is also essential for keeping the immune system healthy. There is also some evidence that low levels of vitamin D may play a role in chronic pain. Vitamin D is typically produced as sunlight is absorbed into the skin and is also put into the body through fortified foods (milk, cereals, etc.), though scientists are currently discovering that many people have lower levels than what is needed to maintain a healthy body. A simple blood test can determine your Vitamin D level (25[OH]D), and a range of 35 to 40 ng/ml is a generally accepted target. (People who live or work in the sun generally have levels between 50-70 nl/ml.)

While too little vitamin D is harmful, too much is very dangerous. Please consult your physician for specific advice.

Swelling under the jaw

An FOP flare-up can occur occasionally underneath the chin and can sometimes be mistaken for mumps, enlarged lymph nodes in the neck or an allergic reaction. It can press up on the base of the tongue, sometimes making it difficult to swallow or breathe. In these cases, this type of flare-up can be potentially life-threatening. To help alleviate swelling and prevent serious complications, a short course of corticosteroids (prednisone) may be considered. Please consult “The Medical Management of Fibrodysplasia Ossificans Progressiva: Current Treatment Considerations,” available at www.ifopa.org or by contacting Dr. Frederick Kaplan or his assistant Kay Rai at the University of Pennsylvania School of Medicine at 215-349-8726. You may also send an e-mail to Kamlesh.Rai@uphs.upenn.edu. The area should not be manipulated, as that may cause more swelling. Special precautions such as elevation of the head of the bed or monitoring may be needed. After the swelling subsides, people are sometimes left with a little knot of bone underneath the chin.

Swelling underneath the chin does not necessarily have to present a major health hazard. Several people have noted hard lumps that appeared underneath their chins which they believe were related to FOP even though they caused no problems.

FOP and the spine

A recently discovered feature of FOP is that the joints in the neck do not seem to form properly in early childhood. Neck joints are formed before birth, but in people with FOP, these joints seem genetically programmed to deteriorate and form bone where cartilage should be. Cartilage is the substance that exists between bones and permits smooth movement of joints. As a result of these abnormal joints, stiffness in the neck may be noticed prior to the observation of extra bone growth. In fact, sometimes babies with FOP won't crawl as a result.

Curiously, the specific problems which occur in the spine also happen in mice that lack a gene called *noggin* that FOP researchers have been investigating in recent years. *Noggin* plays an important role in bone development. Though the *noggin* gene is not the gene that causes FOP, research suggests that people with FOP do not create enough of the *noggin* protein (the *noggin* gene instructs the body to produce a protein that is also called *noggin*) that controls and limits bone growth. Researchers are not yet sure of the significance of this new finding, but answers to what causes spinal deformities in people with FOP likely holds important clues to the condition.

In addition, spinal curvature (scoliosis) may occur in people who have FOP as a result of unequal (asymmetric) bone formation around the spine. In other words, if one side of the body is more restricted than the other, the resulting uneven growth results in an unnatural curve in the spine. Particularly if this happens at an early age, this may restrict normal skeletal growth as the rest of the body continues to grow. Surgical intervention is not recommended because it does not successfully correct the problem and often leads to severe complications, such as flare-ups of FOP in other areas.

Limb swelling

Swelling is a common problem in people with FOP and may result from different causes. First and most common, the limb may swell due to an FOP flare-up. As stated earlier, swelling that is highly localized and nodular is typical in the upper limbs, particularly during childhood flare-ups. In adults or for deeper muscles, swelling may involve the whole limb. This more diffuse swelling is more commonly seen in the lower limbs. While these are typical patterns, either type of swelling can occur at any age.

The restricted movement of people with FOP can also result in a lack of pumping action within the muscle that can cause blood and tissue fluids to pool in the limb. The blood will remain in the muscle instead of being pumped along, and can lead to swelling. In addition, newly-formed bone can press on veins and the lymphatic system, the set of channels which

carry blood and tissue fluid back to the heart. Pressure exerted by extra bone on these vascular channels can obstruct the flow of body fluids and cause swelling. These last two explanations may explain why chronic swelling sometimes occurs in people with FOP.

Finally, and less likely, is the formation of a blood clot. While a blood clot is rare, it is a serious problem. In order to prevent blood clots, it is suggested that the individual wear support stockings and contact a physician to determine if aspirin or a more powerful blood thinner is recommended. When the cause of the swelling is unclear, special tests such as bone scans, ultrasound, CT scans, or MRI scans may be necessary to determine the cause of the swelling so that specific treatment can be prescribed.

If swelling is determined to be unrelated to an ongoing FOP flare-up or a blood clot, a form of treatment that may be helpful is lymphatic drainage. Lymphatic drainage therapy is a type of gentle massage that is performed by therapists who specialize in this area. Lymphatic therapy addresses swelling that occurs when there is more fluid in the lymphatic system than can be moved naturally. The lymphatic system is an elaborate system that does several jobs in the body. Most importantly, it drains fluid from tissues back into the bloodstream and helps fight infection. In people with FOP, this process may not work as efficiently as it could and may contribute to swelling. This is when lymphatic massage may be helpful. If you are interested in this type of therapy, ask your physician for a referral to a lymphedema clinic. (Lymphedema is the name for this type of swelling.)

Broken bones

A fracture in a person who has FOP needs to be treated, just as a broken bone would need to be treated in any person. The goal of treatment in all individuals is to allow the bone to heal in a comfortable and functional position. People who have FOP may not need to have their fractures immobilized for as long as other people, as fractures generally heal rapidly. Surgery is almost never necessary to treat broken bones in people who have FOP. Also, a splint may suffice where a cast may otherwise be necessary. What form of treatment is necessary for a fracture in a person with FOP depends on numerous factors which include: the type of fracture, the bone that is fractured, whether the injury is open or closed, the age of the individual, the degree of deformity, and the functional status of the limb prior to the fracture. The decision on the best type of treatment must be made on a personal basis by the individual's physician.

The extra bones of a person with FOP can also break. If the joint at the fracture site is already locked, no further immobilization may be necessary. Pain medication may be necessary as with any fracture.

Stay comfortable

Sometimes the extra bone created by FOP can make it hard to get comfortable, or a person may find they need extra cushions or pillows to feel fully supported. Fortunately today mattresses and pillows come in many shapes, sizes, and types. With the right solution, it can be much easier to stay relaxed and comfortable.

Here are just a few of the options that are available. As with many aspects of FOP, different solutions work for different people.

- Adjustable beds. Many types of adjustable beds are available. Visit a local mattress store.
- Bucky pillows. Filled with buckwheat hulls. Available through internet and local retailers.
- Gel-filled cushions. Available for wheelchair seats to help with comfort to help with comfort and to help prevent pressure sores. Contact medical supply companies. Hammacher Schlemmer, a U.S.-based catalog-retailer currently sells a “Portable Gel Seat (Item 73077). Call 800-321-1484 or visit hammacherschlemmer.com for more information.
- Microbead pillows. Mogu pillows. Very soft and pliable. Different shapes and sizes. Though Available through internet retailers such as www.amazon.com and www.orangeonions.com. Moosh pillows are available through from www.mooshpillow.com Squishy pillows. Available through internet and local retailers.
- Roho cushions. Roho has been known as a manufacturer of wheelchair cushions for a long time. Now they also make pillows and mattresses. Visit shapefitting.com or call 800-851-851-3449 to learn about Sleepmatterzzz mattresses and pillows. For more information on Roho wheelchair cushions, visit rohoinc.com or call 800-851-3449.
- Tempur-pedic pillows and mattresses. This is a brand name of a type of “memory foam” mattress that conforms to a person’s body. Other brands of memory foam are also available.
- Make your own pillow. One IFOPA member purchased memory foam scraps at a craft store and made her own pillow.
- Also don’t forget to look for solutions in the IFOPA’s *Catalog of FOP Resources*, available at the IFOPA’s website at www.ifopa.org, or by using the tips in Chapter 25, “Finding resources.”

Pressure sores

Skin breakdown and pressure sores are very common and troublesome problems in people who have FOP, especially in adults. Skin breakdown can occur from increased pressure over a normal bony area or an extra piece of bone. Pressure sores can develop rapidly, progress quickly and be difficult to treat. Taking steps to prevent pressure sores is the wisest course of action.

As an article from the Mayo Clinic (www.mayoclinic.com/health/bedsores/DS00570) states, "Pressure sores are easier to prevent than to treat, but that doesn't mean the process is easy or uncomplicated." The following steps should be followed to try to minimize and hopefully avoid serious problems:

- Change your position frequently if you can. If you are in a wheelchair, try shifting position ideally every 15 to 30 minutes. Everyone should shift positions at least once every two hours. If you need assistance, get a family member or caregiver to help.
- Follow these positioning tips. Avoid lying directly on your hipbones. If you lie on your back, support your legs by placing a pillow under your legs from the middle of your calf to your ankles. Avoid placing a pillow directly behind your knees, as this can severely restrict blood flow. Keep your knees and ankles from touching with small pillows or pads. Avoid raising the head of the bed more than 30 degrees in order to prevent sliding, which increases friction between you and the bed.
- Use a pressure-reducing mattress or bed. Options include foam, air, gel and water mattresses.
- If you use a wheelchair, consider one with a tilt feature. Tilting redistributes pressure. Also consider special custom seating if you need to sit for long periods.
- Daily skin inspections are very important.
- Eat a good diet. Healthy eating contributes to healthy skin.

If you detect a pressure sore in an early stage when the skin is red but there is no open sore, it will be much easier to treat. Pressure sores involving open wounds will require more care. Please consult your physician immediately if you detect a problem area, and follow these suggestions:

- Change positions frequently and use special cushions designed to relieve pressure. Avoid pillows and rubber rings, which can actually cause compression and friction and make the problem worse.

- Keep the area clean to prevent infection. A stage one wound (no open skin) can be gently washed with water and mild soap. Anything more serious should be washed with a saline (salt) solution, which you can get at a pharmacy. Avoid using antiseptics such as hydrogen peroxide or iodine, which can damage the skin and delay healing.
- Use a special dressing/bandage that protects wounds and helps promote healing. Name brands of these types of bandages include Tegaderm and Duoderm. These dressings help keep the wound moist (to promote cell growth) while keeping the surrounding tissue dry.
- If necessary, contact a doctor to remove damaged tissue. A wound needs to be free of dead and/or infected tissue to heal properly. There are several ways that this can be done, and a doctor can best determine what to do in a specific case.
- Whirlpool baths, if possible, can help because they help keep the skin clean and naturally remove dead tissue.
- Eat a healthy diet. In particular, vitamin C and zinc can promote wound healing.
- If a wound is not healing, contact your doctor again.

Extra bone below the knee—Osteochondromas

Curiously, approximately 90% of people with FOP seem to have a knob of extra bone on the inside part of the leg bone right below the knee. This bone is nearly always seen in people with FOP even in early childhood before flare-ups begin to affect the knees. This particular type of bone is called an osteochondroma. It generally does not cause any problems. As with the extra bone created by FOP flare-ups, it should not be surgically removed.

Menstrual cycles

A question that women occasionally ask doctors is whether FOP has any effect on a woman's menstrual cycles or periods. Anecdotally, some women with FOP have reported very light menstrual cycles and in some cases no menstrual cycle at all. In addition, some women find that there seems to be a link between their flare-ups and the time surrounding their menstrual cycles. Other women with FOP experience normal menstrual cycles with no unexpected symptoms. Currently it is unknown whether FOP has any effect on a woman's menstrual cycle. As with any case where a person experiences an abnormal menstrual cycle, it is extremely important to see a gynecologist or endocrinologist to determine whether there is a serious medical problem that needs to be addressed. One should not simply assume that a problem is occurring because of FOP. It is also important to note that a woman with FOP can become pregnant, though this is a potentially life-threatening situation to both mother and baby, as the mother's body and organs are already crowded by extra bone.

Does weather seem to affect FOP?

The weather can affect how anyone's body feels. Some people do not feel as comfortable when it is cold or damp. Others are more uncomfortable when it is hot. Still others do not notice any difference with a change of weather or season. People who have FOP do not seem to prefer a particular weather, season, or climate.

14.

Genetics

Genetics is the field of science that studies how traits are passed down from one generation to another. Genetics can easily get very complicated, so we have tried to keep everything as simple as possible.

DNA—The building blocks of us

Each of the cells in a person's body hold very important information called DNA. This information provides instructions needed to develop and direct what happens in our bodies. It determines why some things about us are just like our parents, as well as why each of us is unique. Maybe you are left-handed and no one in your immediate family is, and chances are that there are many things about you that are different from anyone else in the world. That's a good thing—how boring would the world be if everyone was the same!

DNA is grouped into 23 pairs of chromosomes. One chromosome of each pair is inherited from your mother, and the other comes from your father. Each chromosome contains even smaller units called genes. (Altogether, scientists think that the human body contains 20,000-25,000 genes!) As you might have guessed, "genes" is where the word genetics comes from. So genetics is the study of the building blocks that make up us.

The genetic alphabet

DNA is made of four chemical units which form the genetic alphabet. In fact, scientists even use letters of the alphabet (A, T, G, C) to represent the DNA code. Just as the order of letters determines the meaning of words, the order of the letters in the genetic code determines the meaning of the information encoded in that part of the DNA and helps the body know what to do (will your eyes be brown or blue, will you be short or tall, etc.). Arranged in combinations, the four letters contain all of the information needed to build the entire body. A complete set of DNA is referred to as the genome. Think of your genome as a big book of instructions.

If you were to write out the genetic code in the human genome, you would fill a stack of phone books that reached over 500 feet or 170 meters, or about the same height as the Washington Monument in Washington, D.C. If you read aloud the DNA letters that make up the human genome at a rate of one letter per second for eight hours per day, it would take

about a century (100 years) to read. This gives you an idea of how complex the instructions for the human body are.

How a person gets FOP

Most cases of FOP are new, meaning that no one else in the family has FOP. This happens because sometimes unexpected changes (or mutations, the scientific word for change) happen as genes are passed along from each parent. Many of these changes, such as the change that leads to FOP, are accidents of nature that happen for no apparent reason. A small number of cases of FOP are inherited from a parent who has FOP, though this happens rarely because people with FOP seldom have children.

FOP is an autosomal dominant condition. This means that a person who carries one defective copy of the gene that causes FOP will have FOP. Everyone has two copies of each gene, one from each parent. Each of these gene copies, in scientific terms, is called an allele. All of our current information tells us that FOP is *always* a dominant trait. This tells us two things. First, we know that if a person does not show any signs and symptoms of FOP, then he or she does not carry the FOP allele. Secondly, it means that there is a 50% chance that a child of a person with FOP will have FOP too. Let's look at these aspects of FOP in more detail.

Siblings

Parents who do not have FOP should be assured that the chance of having a second child with FOP is rare. FOP is usually a new change (mutation) in the genetic code. As such, for a great majority of families, the chances of having another child with FOP are the same odds of having the first child with FOP, approximately one in two million. Each event is totally independent from the other.

However, researchers have seen at least one family in which parents without FOP had two children who had FOP. In such a family, at least one parent likely had several affected egg or sperm cells. In such a case, the chance of having a second child with FOP is estimated to be about 3% (3 chances in 100). There is currently no way to predict whether this increased risk is present in a specific family.

A sibling or other family member who does not have FOP is no more likely to have a child who has FOP than a person in the general population: that is, one chance in two million. Siblings should be reassured that having a brother or sister who has FOP does not mean that their children will have FOP. To date, no signs of FOP have been observed in the children of unaffected siblings.

Inheriting FOP

Because a person carries two copies of each gene (one from each parent—remember that chromosomes always come in pairs), a person with FOP actually has one “normal” copy of the gene in addition to the damaged copy. As a result, individuals with FOP, both males and females, can pass along either the normal copy of the gene or the damaged gene to a child. If the damaged copy of the gene gets passed along, then the child will have FOP. If the normal gene is passed along, then the child will not have FOP. There is an equal chance of each possibility occurring, so the risk that a person with FOP will have a child who has FOP is 50%.

Pregnancy and FOP

While it may be possible for a woman with FOP to conceive and have children, a pregnancy could be dangerous and life-threatening. The extra bone in the chest, abdomen and pelvis severely limits the ability of the mother’s body, whose vital organs are already overcrowded by extra bone, to adapt to the growth of the baby in the womb. The risk of severe health problems for both the baby and mother is high.

The specific risks include, but are not limited to

- *Risk of FOP flare-ups during pregnancy.* In addition, the use of medications that may help reduce flare-up symptoms may have to be limited.
- *Risk of breathing difficulties during the latter phase of pregnancy.* FOP causes limited expansion of the chest wall, which restricts breathing. As a baby grows in the womb, it presses upward on the diaphragm, one of the major muscles involved in breathing. This further limits that ability of the mother’s lungs to expand. Breathing may be even more difficult if the mother has extra bone that limits the ability of the baby to expand outward from the abdomen. If this is the case, then additional pressure will be created on the diaphragm.
- *Risk of childbirth complications.* Because of physical limitations caused by FOP, a Caesarian section is necessary. Any surgery is a very serious matter for someone with FOP.
- *Risk of general anesthesia for Caesarian delivery.* Caesarian delivery is a surgical procedure that requires anesthesia. Due to FOP, local or regional anesthesia (the type usually used in childbirth) is dangerous and cannot be used. General anesthesia is required. This presents great risk to mother as well as baby.
- *Risk of phlebitis and pulmonary embolism.* Phlebitis is inflammation of a vein. A pulmonary embolism occurs when an artery in the lungs becomes blocked. Both can occur because of blood clots, and both are life-threatening. The chances of

these life-threatening complications are dramatically increased because of the severe immobility caused by FOP. In addition, FOP results in a high-risk pregnancy that requires extended bed rest, further limiting mobility. Lower limb swelling also commonly occurs in the last trimester of pregnancy and further increases the risks of these life-threatening complications.

Specific risks to the child include, but are not limited to

- *Risk that the child may have FOP.* If a parent has FOP, the chance that a child will have it is 50%.
- *Risk of prematurity.* The mother may not be able to carry the baby to full-term because of breathing difficulties. Numerous lifelong consequences can result from premature birth.
- *Risk of severe fetal distress.* Because of the mother's breathing difficulties or other unrecognized problems, the baby may not receive enough oxygen. Because of this complication, there is a risk of death or severe brain injury.
- *Risk of cerebral palsy.* There is a high risk of cerebral palsy due to oxygen deprivation to the baby, especially if fetal distress occurs during the latter part of pregnancy or during delivery. Cerebral palsy is a neurological disorder that affects body movement and muscle coordination.
- *Risk of complications from general anesthesia.* There is a high risk of complications from general anesthesia. More preferable local or regional anesthesia is technically impossible when the mother has FOP.

There are numerous additional concerns. Who will care for the mother during the complications and added stress of pregnancy? Who will care for the child if the mother's physical restrictions leave her unable to do so? And what will be the role of the father, siblings, and grandparents in the care of the child?

Although it is possible for a woman with FOP to carry a child to term, and at least four known instances have been reported in medical literature, pregnancy should be considered very carefully because of the substantial risks to the lives of both mother and baby. Independent genetic counseling to discuss pregnancy and FOP is available if desired.

If a pregnancy occurs, guidance and care at a high-risk pregnancy center is extremely important. At least two lives are at stake: that of the mother and the child. In addition, the lives of other family members will be affected, as by necessity they too will be involved in the consequences of such an occurrence. Pregnancy for a person with FOP has life-altering consequences.

15.

The FOP gene

Scientists have discovered the gene that, when damaged, causes FOP. Learn the implications of finding the gene.

The FOP gene—What does this gene do?

The scientific name of the FOP gene is ACVR1, a gene that is located within chromosome 2. ACVR1 stands for *Activin Receptor Type 1A*. (A receptor is a special protein in the cells of the body that is responsible for relaying information. Some receptors can act as switches that determine whether a particular cell will be a bone cell, muscle cell, blood cell, etc., as well as how the cell interacts with other cells.)

It was only recently discovered that ACVR1 plays an important role in bone development, as well as development of the heart, joints, spine and limbs. The ACVR1 receptor is present in skeletal muscle and connective tissues, although exactly what its normal function in these cells and tissues is not currently understood.

One thing that is known with certainty is that a person can't live without ACVR1. Using mice to test the ACVR1 gene, it has been shown that a mouse embryo that has no functioning copies of the ACVR1 gene cannot develop into a live mouse. (Remember that living things carry two copies of each gene—one from each parent.) In people with FOP one of the copies of ACVR1 is damaged in a very specific way that causes extra bone to form in places where it should not.

Finding the genetic change

The location of the FOP gene was determined by exhaustive DNA research using a small number of multigenerational families in which both a parent and one or more children had FOP. After the gene was discovered, additional DNA testing was conducted on many other people with classic signs of FOP (malformed big toes and progressive extra bone growth) whose blood samples are being saved at the FOP Laboratory. Exactly the same DNA sequence change exists in everyone whose blood was part of this test—in everyone with FOP only one DNA letter out of six billion is different from the standard sequence. (Remember that we discussed that our genetic code is basically a sequence of letters, each of which has a unique meaning.)

The genetic change that occurs in people with FOP is the smallest and most precise change that can occur in a gene. As noted above, one genetic letter out of 6 billion is substituted for another, and that changes the meaning of the genetic instructions. So what effect does this change have?

Human beings are thought to have about 20,000 different genes. While all of these genes are found coded in the DNA of all cells, a particular cell may only activate a certain gene or combination of genes. For example, bone cells and liver cells will use different (though partly overlapping) sets of genes. When a gene is activated, the DNA goes through a process that ultimately leads to the formation of proteins. Proteins perform a wide variety of activities in a cell.

Proteins are composed of a group of 20 different kinds of small molecules called amino acids. In people with FOP, the DNA “misspelling” in the ACVR1 gene causes an amino acid called histidine to replace another amino acid called arginine at a specific location of the ACVR1 protein. To give you an idea of how important this change is, consider that the appearance of arginine in this particular location of the ACVR1 protein has been preserved for all vertebrates (humans, animals, fish, etc.) for nearly 500 million years of evolution. That means that nature has not allowed this particular change because a substitution would likely have severe consequences.

So far researchers have found this same genetic change in ACVR1 in every person with classic FOP. As more people are tested, it is likely that additional changes in the ACVR1 gene will be discovered.

The FOP gene and the future of FOP research

The discovery of the FOP gene validates earlier findings of FOP research that proposed that abnormal regulation of bone morphogenetic proteins, or the master proteins involved in bone growth, seems to lie at the heart of FOP. The gene discovery will also ultimately help us better understand why the molecular switch that produces bone seems to be stuck in the on position for people with FOP and how this process may be influenced by injury or immune system triggers.

The gene discovery will also help us better understand some of the currently unexplainable symptoms of FOP. As previously noted, ACVR1 plays an important role in bone growth. It is extremely important in the development of the hands and feet, both of which can have congenital abnormalities in people with FOP. ACVR1 is also important in the development of the middle ear. As more research is conducted, researchers are likely to determine why

some people with FOP develop hearing loss. Researchers have also recently discovered spinal abnormalities in people with FOP that develop even before extra bone forms. The ACVR1 gene likely holds clues to why this happens as well.

Most importantly, knowing the genetic cause of a disease helps tremendously in attempts to find effective treatments. The discovery of the FOP gene provides the opportunity to produce genetically-engineered mice that have real FOP, a development that would open the door to designing and testing new therapies. While effective treatments will not be available immediately, there is no single discovery that has more suddenly expanded our horizon or given us more hope.

The big question—How long will it take to develop effective treatments for FOP now that the gene has been identified?

This is the most difficult question of all to answer. Truthfully, there is no way to know. There is no doubt that the FOP gene change/mutation is the most valuable piece of information in the FOP puzzle, but it is only the “cornerstone” piece. Researchers still need to understand more about how ACVR1 works—in everyone as well as people with FOP—before they can develop effective treatments.

To develop an effective treatment for FOP, the FOP gene will have to be disabled, blocked, neutralized, or bypassed. Those who work on FOP research have often said that FOP research is like trying to figure out the wiring of an atom bomb so that the bomb can be safely defused before it explodes. The FOP mutation, or the trigger of the atom bomb, is now known. The next step is to determine how to safely deactivate it. This will take time. Development of medications used to treat rare “orphan” diseases is very difficult. Many obstacles can be encountered including issues of safety, drug tolerance, side effects, drug delivery (how to administer a drug, for example pill, liquid, IV, cream, gene therapy, etc.), and determining how well a medication targets the problem. A lot of research and testing must be done. That is the sobering news. But the great news is that we now have an extremely specific target for drug development that will immediately focus an enormous amount of medical and scientific attention on this gene and on FOP.

FOP and other bone conditions

In addition to helping us learn how the catastrophic cycle of extra bone growth that occurs in FOP can be prevented, understanding how the ACVR1 gene works might also one day be harnessed to create bone and skeleton for those who desperately need it. Research

developments may help those who are born without enough bone in their bodies, those who have catastrophic bone loss from severe trauma or amputations, and, of course, for those who have more common conditions such as osteoporosis that affect millions. The discovery of the FOP gene is the most important discovery in the history of FOP research, but it is also an extraordinarily important discovery for all of skeletal biology.

Genetic testing for the FOP mutation

Genetic testing for the mutation that causes FOP is currently being conducted at the Genetic Testing Laboratory at the University of Pennsylvania School of Medicine.

Detailed information can be found at the following website:

www.med.upenn.edu/genetics/core-facs/gdl/

Go to the link for “Diagnostic Tests” for information about the testing process.

For more information, you may also contact the laboratory directly:

Genetic Diagnostic Laboratory
Department of Genetics
University of Pennsylvania School of Medicine
Tel: 215-573-9161
Fax: 215-573-5940

16.

Families meeting the challenges of FOP

We asked ten families to imagine that they were speaking to the parents of a newly diagnosed child. What would they say? Or we asked them to imagine themselves during the diagnosis process. What did they wish someone had said to them? These were their responses in their own words.

As the mother of a 2 1/2 year old daughter, we were absolutely devastated by the diagnosis of FOP. Not only because of what FOP was to deliver to her future, but also due to the fact she had been misdiagnosed and suffered an amputation of her upper right quadrant of her little body.

If we could have fast forwarded our lives from that moment to five years later, we were fortunate enough to find the NORD Foundation (National Organization for Rare Disorders) on the internet, who referred us to Jeannie Peeper, a woman with FOP who was apparently struggling on her own in Florida with the help of her family to start the IFOPA. She was the light in our dark world of loneliness and extreme desperation of FOP. The rest is history.

Going back to the original period of shock having the diagnosis, I can only say it would have been easier on all of us if the FOP association for families had been up and running then for us to ease into the long FOP journey. Once aboard the IFOPA, life became “somewhat normal” once again and sleep came easier, as we realized there was a light burning in a lab in Philadelphia and most probably into late hours, searching for the clue.

Ashley and our family consider ourselves the pioneers of this challenge. We are so gratified to know we have grown extensively and have become a larger family, with still many more to find, and encourage others, that there is life after a diagnosis with FOP.

—Carol Kurpiel, mother of Ashley, 26 years old (born 1981), diagnosed at age 2 1/2



Still relatively new on this FOP journey, I would have to say that the best thing we did so far was to listen to Dr. Kaplan’s brotherly advice to wait and to take our time before learning all we could about FOP. He cautioned us, for instance, that news segments on FOP, though good, are not intended for the benefit of new FOP families, but to elicit an emotional and active response from the general public by showing FOP in its worst and most tragic forms. Dr. Kaplan also encouraged us not to become involved in FOP fundraising right away either. What a relief it was not to feel pressure to do so, as the diagnosis of FOP was a burden enough to handle at the time. So instead, we returned home; we prayed; we cried, and we talked to adult family and friends about Justin’s diagnosis (Justin and his brother, 8, and sister, 9, still don’t know the full extent of his “special bones.”). I soon made some contacts within the FOP community. I joined the FOPonline forum too, though, in retrospect, I think I should’ve waited a bit longer for that, as I found some of the topics overwhelming as a newbie. But, I was warmly embraced, nonetheless, and was blessed to see the resourceful advice, the empathy, the humor, the joy, and the resilient attitudes that FOP families share on a daily basis. Likewise, I also found encouragement in Carol Zapata-Whelan’s book, *Finding Magic Mountain: Life with Five Glorious Kids and a Rogue Gene called FOP*, as it powerfully illustrated to me how, indeed, life goes on (and beautifully so!), and how our family, in our own way, would some day serve as advocates for Justin and the FOP community, as well.

—Wendy and Kevin Henke, parents of Justin, 8 years old (born 2000), diagnosed at age 6



I was born in Belgrade, Serbia in 1979 with the malformation of my big toes. The doctors decided to repair this “flaw of nature” by operating. After a very frustrating period of time that I spent in hospital with my mother, who had to watch her first and only child in pain, I was discharged with stiff toes and no answers. It didn’t even cross our minds that I had a disabling disease which would become a true nightmare nine years later when I was finally diagnosed with FOP after two more operations. Apart from the exact diagnosis, the only thing we were told then was “we don’t know anything about it.” My diagnosis was confirmed in England, where we went on our own initiative, but we learned nothing more there than we already had in Serbia. We learned about FOP from our own experience.

If only somebody had told us about Dr. Kaplan and his team, whom we were able to meet in 1992. He introduced us to the IFOPA and we were finally able to meet other people with FOP in Orlando in 1994, six years after I had been diagnosed. It was quite a shock for all of us. Luckily, we were surrounded by a number of very friendly welcoming people so that about an hour later we already felt that we were part of a big international FOP family.

I wish somebody had told us on the day I was born what the malformation of my big toes meant, and I wish I hadn't had three surgeries. It would have spared my parents and me a lot of tears, painful medical examinations, and fears. Due to my frequent visits to hospitals and all of the things I had experienced in my life, I would start to cry whenever I saw somebody wearing a white coat. I wish that my mother had had other mothers of FOP children to talk to about all the frustrations this disease brings with it.

Finally, I wish I could say there is a way for one to prepare for FOP but, unfortunately, it just bursts into one's life. What I can say is that organizing FOP meetings and making it possible for all FOP families to attend them so that they can share their thoughts, fears and experiences is invaluable for everyone who has to fight with FOP and its cruel unpredictability every day.

—Jelena Milosevic, 28 years old (born 1979), diagnosed at age 9



I simply knew my child would be healthy when she was born. I was the most diligent pregnant woman I could be. So, when Hannah was born with these strange looking toes, and the doctors told me that these things happen sometimes and that the most significant problem she might have was difficulty with her gait and shoes, I accepted these words. I am the middle child of my parent's eight children, so I am accustomed to compromise. For two weeks, I was over the moon.

Then curiosity got me and I did an online search for toe deformities. I was directed to the FOP website, and, as I sat there and read, a sense of panic overtook me. I pushed the fear to the back of my mind and hoped and prayed that maybe, just maybe, Hannah didn't have FOP. Months passed. However, when she was eighteen months old, she developed her first swelling. I knew. Deep down in my heart, I knew. However, I went through the formality of taking her to the pediatrician and stayed in a state of denial until someone mentioned the word biopsy. I remembered the website information retelling of explosive episodes of bone

growth from invasive procedures. Compelled by my fear of Hannah being hurt, I asked the doctor if he had heard of FOP. He narrowed his eyes and frowned at me. I felt stupid, but I stood my ground to protect this little person whom I loved more than myself.

Hannah is now six years old. We have been dealing with FOP for five years now. Some part of me has cried, worried and pleaded with God, asking why my child has to hurt in the middle of the night and why the simplest task is an impossibility for her. However, the part of me that loves my little girl, still more than myself, today more than yesterday, thanks that same God for allowing me the opportunity to take care of her and love her as she deserves to be loved. Hannah has a health problem, but she is also a miracle to me. She is that thing, that special person who makes it possible for me to better understand and appreciate the importance of a smile, a kind word, an understanding gesture. Dealing with being the parent of a child with FOP is a gradual, ongoing process. I softened gradually, accepted gradually, learned in small increments. I am still learning. I still get angry, depressed and sad. But my prevailing thought is love. As Hannah's mother, I try to make every day more special and bearable than it might have otherwise been had I not been involved. I forego selfish pity and laugh and smile with my child. The return on these small gestures is more valuable than gold, and our children deserve nothing less.

—Sharon Davis, Mother of Hannah, 6 years old (born 2001), diagnosed at age 18 months



Your child has been diagnosed with FOP. This is not the end of the world. You will recognize that it keeps the family together. It is also important that your child grows up as a normal child. You should inform your family, friends, kindergarten and school about FOP and give them information about FOP to help them learn more. Over the last 19 years the FOP community has grown a lot. You will find information about FOP at different websites, with reports, different educational materials, and a lot more. There is a chat group where you can ask what you would like to know about FOP. You can ask different people with FOP about their experiences and how they found helpful items to cope with the loss of mobility.

In Philadelphia there is a laboratory devoted to FOP. Since April 2006, we know what the FOP gene is thanks to the people who work there, and they are working very hard to find a cure. Life is beautiful and together we will find a cure!

—Roger zum Felde, 42 years old (born 1965), diagnosed at age 2 1/2



When my daughter Jasmin was diagnosed with FOP, we had just moved from Wisconsin to Connecticut. I was very lost and alone and wished I had a friend or family member to hug me or help me. Neighbors tried to encourage me and tell me that it would be okay, but it wasn't. It was as if I was in a nightmare that never went away. After some time, we found Dr. Kaplan and the IFOPA, and I gradually began to read the information packet that was sent to us. But it was definitely way too scary and too much to process. Thankfully within a couple of years a doctor from the Hole in the Wall Gang Camp for children with illnesses and disabilities got us set up with a whole team of specialists, including a social worker, a pulmonologist, and a pain team. This provided us with an important framework and continuity of care. We no longer felt as if we had fallen through the cracks in the system. FOP is still not easy to deal with sometimes, but I have learned some things along the way. First, try to appreciate the little moments that happen every day and enjoy the spirit of your child. Take each day as it comes. Try to fear less and love more. Another thing that helped me was jumping into the IFOPA through advocacy, fundraising, sharing information, and raising awareness of FOP. These things really helped lessen my feelings of hopelessness. I think it is also important to never give up and to always have hope in your heart.

—RoJeanne Doege-Floyd, mother of Jasmin, 13 years old (born 1993), diagnosed at age 5



I have been followed by people who think I shouldn't be carrying groceries. I am often approached on the street and asked if I'm okay. Once, a woman came up to tell me that what I was doing was "inspiring." Smiling, I bit down on my tongue to stop the words "What, walking?" from coming out of my mouth.

I tell of all this to illustrate how idiotic (or, if you want to be nice, naive) the world can be. The belief is that if you happen to be born with a disability, you are sentenced to a second-class life. This is because people have become set in their ways, utterly convinced that there is only one way to do things.

What's been forgotten is that we, as mammals, are warm-blooded creatures. We're meant to adapt. It is because of this selective amnesia that I spend my life battling with people who consider me helpless. (I'm not "able-bodied", how can I do anything?)

Luckily, I figured out rather early on that, seeing as they have never lived in my body, people know very little about what it can do. Thus, these folk (often in mid-stride as they rush to the damsel's aid) end up looking on in disbelief as I manage without them. (FYI: the squeeze handles on bottles of cleaners are perfect for getting cans down in grocery stores). On the plus side, their confused looks are rather amusing. The negative? It's going to take me a long time to pull down cans in front of every person who thinks I'm helpless.

The sad thing is that people who remain in their one-way rut will never know the joy that comes from figuring out new ways to do things. My way will not be your way, but if you look past the conventional, you might find that you can do more than you thought.

—Marin Wallace, 26 years old (born 1981), diagnosed at 3 1/2



The best advice we received (and have been guided by ever since) was from Jeannie Peeper and Val Pinder, both adults with FOP who stressed letting Oliver lead a normal life, as the FOP was going to progress and no amount of protection would be able to prevent this. As a result, Oliver has enjoyed a very normal, relatively unrestricted childhood (apart from not playing contact sports in a formal sense) and has grown into a very well adjusted, confident, contented young man despite the progression of the FOP in his teenage years. Val stressed the importance of education, as the FOP could not affect his brain, so he has done a lot of speech and drama and debating, music etc, which has helped his confidence and ability to speak in public. Oliver has been empowered from when he was quite young to be involved in the decision-making in relation to the FOP and its implications. In consultation with his doctor or teaching staff and parents, he decides what activities he will engage in, when to take extra pain relief, when adjustments are needed in class and how to tackle awareness raising when it has been needed in the past at school. He has always had knowledge of FOP relevant to his age and level of understanding, but it has not been allowed to dominate his or our lives—although it is always there at the back of my mind as a mother! Through his many activities he has made some wonderful friends, and because he has openly discussed and hence demystified the FOP at school on many formally organised opportunities, he has enjoyed wonderful support from staff and students alike and genuine friendships have evolved. So my advice would be to follow the advice we were given because, to quote another parent of a child with FOP whose name escapes me, “Your child need not be emotionally ‘crippled’ just because they are physically affected.”

—Julie Collins, mother of Ollie, 14 years old (born 1993), diagnosed at age 18 months



As someone with FOP, I realize how the first reaction as a parent is to protect a child as much as possible. But you also need to think of your child's life and not restrict your son or daughter too much. I know that as I was growing up my parents thought they were doing their best, but I missed out on so much because they were afraid and wouldn't let me attempt new things. You need to find a good balance between safety and allowing the child to be...well, a child. As parents, simply use common sense...let your child play with friends... don't let your child play contact sports...and ask your child what he needs and feels up to doing. Growing up, I felt out of control of my life and even more so when FOP started because I lost a lot of my choices. My parents were over protective and I missed out on part of being a kid as a result. Your child may surprise you by telling you that he knows his limits, for example by not doing something that he can't or shouldn't do. Just be supportive. We with FOP are a stubborn lot. Just keep an eye on your child and watch to make sure he isn't pushing too hard just to feel normal. Above all, make sure the child is happy and knows you love him and have only his best interest and future in mind.

Jonathan Carmichael, 30 years old (born 1977), diagnosed at age 9 (FOP symptoms started at age 7)



It may not be evident now but your child has been chosen to change the lives of others, as well as your own. You will experience many different emotions such as denial, anger, sadness, hopelessness and joy as well. No one can possibly comprehend the sadness you are feeling now, but please accept the words of comfort that people offer you. When our son Cody was first diagnosed, it seemed like a bad dream. We could not imagine how FOP could be real and how it could have happened to our child, our family. We felt as though we were alone and no amount of cards, meals or prayers would help ease our pain. Friends and family wanted to understand what our family was going through. They desperately wanted to say the right thing but didn't know how. In time, we soon felt *God's* blessings and realized we were not alone on our FOP journey, and neither are you.

There are people in this world that know exactly what your family is going through right now. They are mothers, fathers, grandparents, cousins, aunts, uncles, doctors, scientists and a

group of special people that are just like your child. These special people were diagnosed with FOP and are the strongest, most well-adjusted people that you will ever meet. Most are willing to share their life experiences. I could not have gotten through that first year without the friends I met through IFOPA and FOPonline.

Some say it is more tumultuous for the parents than the child who is diagnosed with FOP. Everyone is given obstacles in life that they must overcome. Embrace this obstacle as a gift and don't let it all be about FOP. You have a child with a rare condition but he/she has other strengths that you need to focus on, especially now. You may also have other children. Don't push them aside and ignore them or their feelings. Their lives will be affected by this also. Let them express their feelings and concerns to you.

My advice is to live each day to the fullest and take it one day at a time. When your child was born, you held him/her in your arms and you imagined their future. I know that you never imagined a future so frightening as this, neither did I. Please have hope and seek comfort from your new friends in the FOP community. You will find comfort in knowing that you are not alone. Have faith that there will be a cure for FOP. Until then stay positive, for your child as well as for yourself. Never give up hope.

I believe that each child with FOP will have a special place in heaven. There is a reason for everything. I have searched for answers as to why my child was chosen to have FOP. I have found some comfort in the Bible. One of my favorite quotes comes from James 1:2-3 in the New Testament: "Consider it pure joy whenever you face trials of many kinds, because you know that the testing of your faith develops perseverance." It is true—FOP does make you stronger. In time, people will tell you how strong you are and how inspiring your child is. Soon you will realize that your child has been given a mission in a life. This mission is to show others how to live life to the fullest.

"The greatest light is always concealed in the darkest covering."

—Michael Berg, *Blessings and Light*,

—Jen Dennings, mother of Cody, 12 years old (born 1995), diagnosed at age 8



A note: In this chapter and the chapters that follow, ages of individuals are the ages at the time when each piece was written (in some cases as far back as January 2007). A conscious decision was made to do this in order to preserve life and feelings in the present moment.

The IFOPA's religion policy

The International FOP Association (IFOPA) is a non-denominational/non-religious organization, and, as such, it does not endorse, serve, or favor any specific religious organization, practice, sect, or idea of any kind. The information below is that of the author's opinion and is included in this guidebook as an individual's personal story.

17.

Finding magic mountain: *Our family's life with FOP*

by Carol Zapata-Whelan

My son Vincent was diagnosed with FOP in 1995, when he was nine years old. His first symptom was a mysterious limp, which, thanks to careful, intuitive specialists, led relatively quickly to a diagnosis without invasive tests. It is also by luck, magic, and miracle that we found Dr. Kaplan through a book in a bookstore, before the telepathy of the internet. Since 1995, our family has been through myriad trials battling FOP and we continue to battle it, hoping, praying for a cure. I am so proud of Vincent, who looks for ways around FOP to reach his goals. Vincent represents just one more example of the courage and perseverance that defines the FOP community. On August 8, 2008, he will give a speech about life with FOP at a White Coat Ceremony as he is initiated into the University of California, Irvine's School of Medicine. Vincent hopes to one day—somehow, some way—help Dr. Kaplan find a cure for FOP.

In my family's own difficult journey with FOP we have experienced small and large triumphs and miracles and learned vital lessons. I have found that when we cope with a child's mysterious disease, we look for information, doctors, treatments; we do all we can to help with school, friends, and special needs. But one thing we cannot always do for our children is take away suffering. Then, I believe, the challenge is to find the magic from the mountain we climb together. This image of a mountain is important to me because I entitled a book about our journey to raise FOP awareness *Finding Magic Mountain: Life with Five Glorious Kids and a Rogue Gene Called FOP*. A mountain can represent both a great obstacle and a great vantage point from which to discover the undiscovered. It is from this memoir that I share a few perspectives below. (Coincidentally, after the book was published, the 2007 FOP Symposium had as its theme, "Together We Can Move Mountains"—so a even mountain is not immovable!)

When Vincent had his second major FOP flare-up in 1997, there was nothing I could do to take away his suffering, to stop FOP. Desperate, I wrote to a healer across the sea. I sent her a photograph of my son in his Catholic school uniform. Not long after, the healer called. She said she had seen my son's face in a dream. She told me she had seen a soul pass through a flame, which signified suffering. She then explained that FOP came from an

ancestral curse. At first I was confused: FOP is such a mysterious ailment. How else could anyone explain it? And of course as a mother, I felt that FOP was somehow my fault. A geneticist at the University of California San Francisco had already pointed out—though incorrectly—that FOP came from my side. Disoriented, I listened to the healer and finally thanked her for calling. But after hanging up, I was very certain of one thing: FOP is no curse—no disease, no earthly challenge is a curse (never mind that I don't believe in such things). Instead it is a *course*, a path to follow up a mountain, one which can—strangely—strengthen and lead to unimaginably dazzling people, places and dreams. To give you an idea of this course, I would like to include the words of others in the FOP community, friends from around the world who have also shared their pain, courage, wisdom, hope and joy. This community has come about thanks to one indomitable woman, Jeannie Peeper, an FOP adult who—through most of her life—had never met another soul with FOP. Years ago, Jeannie reached out with a letter to another FOP patient and started a vital support network that connects families around the world and raises hundreds of thousands of dollars for research and special needs assistance. All of the friends whose words I share with you today I know because of Jeannie Peeper.

The night I spoke to the healer in the Philippines, I was worried about my son and mystified by FOP. When we are faced with a difficult diagnosis, I think we are at first confused and ask ourselves questions with no answers. Why did this happen? How did this happen? Why my child? These questions were behind the words of another FOP mother in New York, Connie Green, as Connie wrote in a letter:

When Sophia started her first body flare-up, I walked around contacting everyone who knew anything about medicine, FOP, Sophia, me...During that time, I thought I was grounded in my body, but I was not present. Stress does things to us that we can hardly be prepared for. And FOP is such an unusual, isolating stressor that my mind had gone into hyperdrive to find some of life's sweetness and normalcy, to escape the pain and yet find a way to accept that Sophia and I had been captured by this enemy and given a life sentence, having committed no crime.

I think that maybe we come to accept a challenge like FOP in different stages: one day, yes, we accept it—and one day, no, we do not, moving back and forth until acceptance sets in for the most part and life becomes routine again. When Vincent was nine years old, we explained to him what we believed he could understand: that a bone could grow in his muscle if he hurt himself, so he had to be careful; it would be too dangerous to ride a skateboard, for example, or to play football. We did not explain too much—just what we felt Vincent could understand at his age, and what he needed to know to keep safe without

impossible restrictions—and we always made sure to tell him that there was great hope in Dr. Kaplan’s research and that God would take care of everything.

I do believe that our children begin to accept difficult truths even before we can explain them—in ways we do not know, and on their own schedules. When I say this, I remember a conversation I had with my daughter, Celine, who happened to be four years old when we were deciding whether or not to take Vincent to his first FOP family meeting. We fretted over whether or not he would be OK, at the age of ten, meeting grown-ups immobilized with advanced cases of his condition. Interestingly, the conversation I had with my four year old shed a light on how children might begin to process difficult truths, and I include this exchange here from our memoir:

One day I took a stroll with four-year-old Celine. She rode her bike with training wheels, and I walked alongside. On our way, I spotted a lifeless tabby cat in the bike lane, its eyes open and glassy. To shield her from the sight, I walked between Celine and the animal, distracting her with stories and questions as we went.

On the way back, Celine saw the lifeless cat before I had a chance to shield her again. “Look!” she said, stopping her bike.

“Yes, the poor cat’s dead,” I explained.

“But its eyes are open,” said Celine.

“It’s still dead.” The animal looked intact, probably sideswiped by a car.

Celine studied the tabby for a while. “Why can’t we have a cat?” she asked, finally moving on.

“I’m allergic to cats.”

“How about if we just get a dead cat?” Celine’s blue-green eyes looked so earnest as she pedaled. She made it sound like a perfectly reasonable request.

“What would we do with a dead cat?”

“We could look at it,” said Celine, “and then we could bury it.” We buried Teacher Blanche’s cat in her garden one day.” Celine’s eighty-year-old preschool teacher had not shielded her little students from this loss.

I realized that though Celine might be too young to fathom the reality of death, she was not too young to start to accept that life held loss in different forms. In some ways, I thought, meeting loss in FOP and accepting the possibility of an unreal future loss might not be so different. I had tried to shield

Celine from a dead cat that she ended up accepting so naturally, she even suggested we get one for ourselves. Even if she was thinking as a four-year-old thinks, Celine seemed able to process the understanding of loss.

After my walk with Celine, we simply went ahead and asked Vincent how he felt about attending the upcoming meeting of FOP families. These gatherings have been precious vehicles to unity and solidarity, unique opportunities to hug old friends one only knows by phone or e-mail. One such friend—my most frequent FOP “advisor”—is Sharon Kantanie. Sharon has made monumental contributions to the IFOPA with her writing and organizational talents. Like Jeannie Peeper, Sharon has been a shining model for our family; she has guided me through flare-ups, medications and college transitions. Sharon’s practical advice, words of comfort, wisdom, and encouragement have made all the difference. It was at an FOP gathering that I met Sharon. As for a visit to our first IFOPA event, Vincent responded that he would go with us to see Dr. Kaplan, but that he would meet everyone at another reunion. And this answer was perfectly fine. We would face loss one child at a time, one event at a time, one generation at a time, in the best way for each child. A few years later, Vincent was overjoyed to meet everyone, and that IFOPA symposium contributed to the impetus and inspiration for his deciding to become a doctor.

When FOP first came into our lives, one of the hardest challenges for Vincent was having to give up activities that could cause trauma. There were times when Vincent’s friends would play a game that was dangerous for him and we could not allow him to take part. It was heartbreaking to not be able to answer when he asked, “Why me?” But it was also vitally necessary to allow our son to live actively. We worked together with his school and an adaptive PE teacher, an occupational therapist, and a psychologist to establish safe recess games for Vincent—to adapt sports so that he could still have fun, but not unduly risk his mobility. These substitutions, of course, were not easy, and Vincent missed being allowed to ride a skateboard. But what was especially important is that we discovered wonderful alternate interests, in particular, music. In high school Vincent was in the marching band at school. In college, he played in a university orchestra thanks to the trumpet and the piano his father made sure he learned to play. When Vincent could no longer take part in organized sports, his father made sure he had plenty of other activities, with music at the top of the list. In one of his college application essays he wrote, “I owe my love of music to my father.”

Life does go to a “new normal” as FOP mother Marilyn Hair, who has long served the IFOPA, aptly describes it. And then the “new normal” may change in other ways. After Vincent began music and our adaptive PE teacher saved the day, I was happy that we had “fixed” our problems at school. But one evening at dinner, when Vincent was fourteen, he was remembering school recesses and suddenly became very angry that other children had

left him out of their usual games. He rose violently from his chair and slammed the back of it against the table before stomping off. We sat in silence around our unfinished meal, not knowing what to say. “It’s the prednisone,” I finally explained. Prednisone can affect moods, as many of us know. “When he’s off prednisone, things will be better,” I said. Just then Vincent’s younger brother Lucas, who has always shared a room with Vincent, quietly pointed out, “Yes, but he still feels that. The difference is the prednisone lets him say it.” Our then eleven-year-old son Lucas understood what I did not until that day: that you can adapt and “fix” things for your child, but you cannot always “fix” the people around you and that there are certain feelings that you cannot always “fix.” For these instances I remember the words of FOP mom, Jeri Licht in New York, who learned this lesson well before I did as she wrote one day:

When I was pregnant, I was very panicky about the responsibilities before me, even though I had never yet heard of FOP. A psychologist told me that kids are not as breakable as I feared. She said the words “don’t know” and “I’m sorry” will go a long way to smoothing the road. I think that comment and those phrases have saved me. When I can’t think of anything positive or hopeful to say to Daniel about something, I say, “I don’t know.” When I overreact or Daniel complains about FOP, I say, “I’m sorry.” Those words have certainly come in handy.

On our way up the mountain with FOP one especially important direction we have taken has been finding caring medical professionals—and toward helping them understand FOP. Along this path I have learned that in the face of something complicated, a sense of humor can make all the difference. With FOP, for example, we must usually tell other doctors or nurses what they can and cannot do, as they have never seen a case of the condition. I have found that it helps to define FOP, to give clear accounts about what is going on, to stress warnings about what can and can’t be done, and to have notes and questions ready. To illustrate, I relay a memory of the time Vincent allowed medical residents at a conference to try and diagnose him from clinical findings. As usual, I had to warn everyone that in FOP passive exercise is forbidden:

Two young men and three young women: they look harmless enough, and one of the men, teddy-bearish, with a mustache and glasses, seems the friendliest. I give them all *The Warning*—as hastily as possible—“Do not push his limbs past capacity. My son’s arms, neck, legs must not go past their

“give,” no passive exercise; please be careful.” Their intelligent eyes are all on me. They, Vincent, myself, are all much too serious. So in conclusion, it occurs to me to add, “Or I’ll kill you.”

Even the teddy-bearish resident looks over a little sharply, unsure...[But] Vincent breaks into a smile...and the least tired looking of the group laugh. The ice is broken. So for the rest of the hour, I will hear myself repeating amiably to each team in rotation, “If you try to move his limbs past capacity, I’ll kill you.” A nurse accompanying one of the groups greets me cheerfully: “I heard you telling people you were going to kill them—so I was waiting to find out what not to do!”

So many of the lessons about how to live life with FOP I have learned from other mothers. Another mother in Sweden, Marie Hallbert, often shares her stories. Years ago, she wrote words that have stayed with me for a long time, and that exemplify many people I know who overcome FOP daily in their lives, whether parents or children:

I just want to share a little story. Six years ago I read an interview with a famous Swedish girl name Kristin. At that moment she was pregnant and she got the question, “Are you worried about if your child is having an illness in some way?”

Kristin answered: “Once an old wise woman said to me, ‘If you get a child with a disease, you will have that child because you have got the strength to handle that.’”

And I had those words in my head when I was pregnant with Hugo. And when we got the diagnosis two years ago, I had the feeling that I must be a very strong and special person to get a child with a disease like FOP. There must be something meant for me to do. And I have the same feelings about Hugo. Even if life feels like shit (sorry about the expression) sometimes, too.

I have seen small and large miracles take place—magical moments—because of the resolve and courage of FOP families. And here I come to one of the small miracles of the trek up this mountain. Moira Liljeström, a mother in Argentina, resolved to raise FOP awareness through the press. Like Marie, she also helped found an FOP organization in her country. Moira’s sense of purpose led a young girl in Argentina to a correct FOP diagnosis. And miraculously, Moira helped find a multigenerational FOP family in Korea—there are

only seven such families in the world—which allowed FOP research to advance more quickly than anyone might have imagined . A miracle took place thanks to Moira’s persistence: The location of the multigenerational FOP family in Korea was one of the last pieces of the puzzle that Dr. Kaplan and Dr. Eileen Shore with Meiqi Xu and other members of their research team needed to discover the location of the FOP gene two years ago. Moira wrote to me in a letter one day:

One of the tasks we set for ourselves was to look for and make contact with FOP families in Argentina, as communication is the best way to face FOP. And after reading about the hunt for multigenerational FOP families to help DNA research, we realized that this search was a task that we could carry out in our own country. We found ways to spread information in popular and medical publications. Sharing information was a way to help others avoid the long and difficult process of arriving at a correct diagnosis. In 2004, we helped Dr. Kaplan find a multigenerational FOP family in Korea, and a fifteen-year-old girl in Argentina correctly diagnosed herself when she read the first article that appeared on FOP in one of our national newspapers, *El Clarín*.

I have found that the solidarity of the FOP community has produced other miracles too, miracles that are like “magical” coincidences—yet they are not coincidence—I choose to see them as small miracles made possible by faith, prayer, persistence and by the families and medical professionals who labor relentlessly. The following is the story of Kelly Alexy, a nurse-practioner at the University of California San Francisco, who helped diagnose a toddler with FOP after connecting a few vital dots. First, her sister, Vincent’s science teacher, attended an FOP fundraiser and then asked Kelly if she had ever heard of the rare condition.. Kelly later learned that her superior, UCSF neonatologist Dr. Joseph Kitterman, had a grandson with FOP. Dr. Kitterman also invited Dr. Kaplan to speak at UCSF. Here is an excerpt from a letter that Kelly sent my family one day:

I was on service in the newborn intensive care unit as the neonatal nurse-practitioner. Three times a week we would go to the radiology department to look at MRIs, X-rays, and ultrasounds of the babies in the unit. Sometimes we would have to wait while other departments looked at studies of their patients. We were waiting for the neuroradiologist to finish discussing a study that was done of a growth on the neck of

what I knew to be a two-year-old. The patient was being followed by the hematology/oncology department.

There was an open discussion within the group of doctors and suggestions regarding infections and asking if biopsies were made. The oncologist, Dr. Goldsby, mentioned that a biopsy was done and was negative and that the growth had migrated down the spine and the child had decreased mobility where the growths were. I did not say anything at first, but I mentioned to the neonatal fellow that it sounded as if it could be that disease that Dr. Kitterman's grandson has....

...I was not going to say anything aloud to the whole group of doctors, so when Dr. Goldsby was done and was leaving the room, I tapped him on the shoulder to see if I could ask him a question regarding his patient.

He said "sure" and I asked if the child had normal toes. With a slight look of surprise he said no, in fact, they had just noticed that morning that the child had short great toes. He then eagerly asked me why I asked that. I told him that I knew of a very rare disease where children get swellings that then ossify, which is really hard to diagnose. These children are referred to oncologists and have biopsy after biopsy and even get treated with chemotherapy. He asked me the name of the disease.

He said that he was willing to investigate anything because this child's condition was so perplexing. I said it was called FOP. I asked a colleague if she recalled the name of the doctor from Philadelphia who came to UCSF to lecture on the disease. She gave me Dr. Kaplan's name, and I called Dr. Goldsby and gave him the information.

Thank goodness the events happened as they did. It is quite a coincidence. I think my chances of winning the lottery are better than me walking in on a conversation of doctors discussing an undiagnosed child with FOP. I am so happy this child did not have to go through any more unnecessary testing. It makes you think sometimes you are put in a certain place for a certain reason. I often wonder if in my life there have been perfect strangers whose actions have changed the way my life is lived.

Dr. Kaplan responded to Kelly's story with a note of his own: "The bad news is that [this little one] has FOP. The good news is that he has a guardian angel—Kelly Alexy."

It is also important to mention another miracle on this climb up the mountain following FOP. Dr. Joseph Kitterman, who helped make the diagnosis of the young boy possible, founded the world's second FOP Center at the University of California San Francisco in 2005. The San Francisco Chronicle announced this news on its front page in March 2005. Vincent also had something important to do with the creation of this center and he was its first FOP patient.

Our loved ones facing a health condition like FOP can make our lives more challenging, but challenges like FOP can also inspire the best from the world and from our families. When Vincent began his studies away from home at the university, this was a difficult time for his parents, far away, and he counted on his brother Brian for anything necessary, pharmacy trips, help fixing the medical scooter, moving furniture, and even intervention with an graduate student instructor who accused Vincent of losing his work. When I asked Brian how he got a graduate student instructor to allow Vincent full credit, for the lost work, he said, "I told him I was bigger than he was." So from our oldest to our youngest our family has faced FOP together, climbing this mountain together. When our youngest, Isabel was eight years old, she founded a club, The Best Friends Forever FOP Club, in which Dr. Kaplan is a proud charter member (he sent the very first club dues). The rules for Isabel's club hang in the hallway of the University of Pennsylvania's Medical School. They state:

- I.
 1. Ideas are powerful things.
 2. Ideas are like inventions.
 3. Share your ideas;

- II.
 1. Pay attention.
 2. Respect other people's ideas.
 3. Feel free to ask questions or share any ideas.
 4. Listen to other people's questions.
 5. Be creative.
 6. Wait 'til it is your turn.

(These are rules forged through trial and error by five glorious kids.)

Finally, I would like to quote the beautiful words of joy and gratitude of Norbert Seidl, a young German with FOP who told Dr. Kaplan of how he thanked his mother in a

very special way for his life. These words express the joy in life that a challenge like FOP cannot take, but can strengthen in those who climb a mountain with love, faith, hope and solidarity:

We have had FOP our entire lives. We know our FOP may get worse, but our joy in life is not affected by this fact...As we have grown older, our needs have increased...All along, our mother has been there for us, to help us and to help make our lives better. One day, I said to my sister Christine, "Our mother deserves a medal for all she does for us every day. It is important, at least once, to say thank you to our mother in a very public way. There are millions of people who live in Germany, but our family is unique. We are the only multigenerational family with FOP in all of Germany...So I wrote to the President and I told him the story of our family...I told him that our mother deserved a medal for all she did for us every day...and I also described how she looked after our father as well, who also had FOP. The President was convinced, and he decided to award this historic medal to our mother. A letter arrived one day in the mail from Berlin, and I, the mailman, delivered the news to our mother. The letter said that our mother would be awarded the highest civilian honor of the German government...We all attended a beautiful ceremony at a castle in Bavaria, where the Secretary of State... presented the medal to our mother.

All those I have met in the FOP community, families, children, parents, medical professionals, share in the precious medal Norbert Seidl obtained for his beloved mother.

Years ago when I spoke to a healer, guilty over whatever my role in our son's suffering might be, I was told a curse, a sin from an ancestor, was responsible. Of course, those past lives, those ancestors in question, can only have been Adam and Eve, parents of the human condition. As our family makes its journey on the steep course tracing the path of FOP, I am learning that life is a magic mountain rising over fields of dreams for as far as our eyes will see.

The above article is a version of speeches my son Vincent and I gave at Cheng Kung University Hospital in Taiwan, Taiwan. At the invitation and because of the generosity of the Taiwan Foundation for Rare Disorders, we visited this hospital in May of 2008 with Dr. Fred Kaplan, who gave an inspirational lecture. In Taiwan we formed immediate friendships with FOP

families and caring medical staff. At the hospital we also met an Asian film star, Jessie Chang, who told us that she had just finished making a movie about FOP and that she and the little boy of ten who played her son wished to meet a young man with FOP. We were astonished to learn that the first major feature film about FOP would appear in Taiwan and cross the Taiwan Strait to China. We were even more astonished to learn that the film came about because a Taiwanese movie director read Finding Magic Mountain in Mandarin. Miracles happen. The next one, we pray, will be the miracle of a cure for FOP.

About the author

Carol Zapata-Whelan, raised in Argentina and the United States, has a doctorate in comparative literature from UCLA. She teaches at California State University, Fresno, and is the proud mother of five children. Her writings to raise FOP awareness have appeared in Newsweek, Hispanic Link News Service-Los Angeles Times News Syndicate), Chicken Soup for the Latino Soul, El Andar, The Rotarian, and other places. Her book (to draw attention to FOP research and patient needs) Finding Magic Mountain: Life with Five Glorious Kids and a Rogue Gene Called FOP has been translated to Mandarin and Korean.

The IFOPA's religion policy

The International FOP Association (IFOPA) is a non-denominational/non-religious organization and, as such, it does not endorse, serve, or favor any specific religious organization, practice, sect, or idea of any kind. The information below is that of the author's opinion and is included in this guidebook as an individual's personal story.

18.

My children

by Dorothy Kadala

When Sharon Kantanie asked me to write about siblings and FOP, I wasn't sure I could think of anything useful to say. Added to that is the fact that they are all grown now, and my memory is not what it used to be. So... I'll just begin at the beginning. Susan was two, almost three years old when we first knew that there was something very wrong with her. At that time we had Gerich, age seven, and Ann, who was 7 months old. We lived in The Netherlands, far from family. We were fortunate to have good friends, both American and Dutch, who were very supportive. However, from the beginning Gerich began to take on tasks that belonged to an adult. He was very helpful at home and was indispensable for running errands around our small village. He went to the bakery, the butcher shop, the grocery store, and other places for us. He was still an active little boy who loved school and playing with his American and Dutch friends.

Neil arrived about 18 months later. Shortly after his birth we moved to a new area of The Netherlands, far from family and friends. It was not long before the children had a wide circle of friends. We tried sending Gerich to the closest international school, but the two hour bus ride was just too much. We enrolled him in the local school, just minutes from our house, as Gerich could already speak and read Dutch. That opened up the neighborhood children to us. Susan traveled about 45 minutes to a school for the disabled. Gerich continued to be the helpful big brother. I tried to make life as normal as possible, but he and Ann were used to Phil or me heading off to the hospital for days at a time. I usually took the babies with me when I took Susan to the hospital (first Ann and later Neil). I was breastfeeding and the hospital personnel were quite accommodating.

After two years of battling what turned out to be a misdiagnosis, we returned to the United States, specifically Wilmington, NC. (with a nine month detour with my mother awaiting Phil's transfer). Susan was correctly diagnosed with FOP by doctors at the Medical University Children's Hospital in Charleston, SC when she was six years old. At this time Gerich was 12, Susan 7, Ann 5, and Neil 3. We spent the next 17 years in our house on Bayshore Drive. The children attended the local schools and after a year I returned to my job as a school librarian. It may seem strange, but our lives seemed very normal. Our children went to school, to church, Scouts, etc. They took piano lessons and played soccer and other sports. Susan could not participate in all those activities, but whenever possible we attempted to let her do things. She had a reclining three wheel bike called the "banana peel" and she

raced around the neighborhood on it. She had many friends from the neighborhood, church, and school. We tried to not make her feel different. She had to do her school homework, and she had chores to do just like her siblings. They were things that she was able to do. Sometimes there were complaints of unfairness. And “why doesn’t Susan have to do....” Sometimes we had to have family meetings to remind Gerich, Ann and Neil that Susan could not do all the things that they could do. I always told them that we were a team. Each member of the team had responsibilities. All jobs are important, and our family could not make it without everyone doing his or her job. They were so used to the way Susan was that sometimes they had to be reminded. Gerich was older and involved with middle and later high school activities and friends. He was a great baby-sitter (sometimes he begrudged it, but in the main he enjoyed the income). Sometimes we had help with housekeeping, but as the children got older they took on many of the housekeeping jobs. By the time Gerich reached high school, Ann had become Susan’s and my biggest helper. This was particularly true on the bus and at school. As a sister, Ann was and is able to help Susan with bathing, dressing and toileting, things a brother is not likely to do. Until Susan got her driver’s license, Gerich and later Ann drove her places. Susan is no longer able to drive, so once again her siblings take her places when they are home. Ann invites Susan to visit her in Chapel Hill for weekends to give her and us a break.

We did not limit the other children from doing things that Susan could not do. Gerich and Ann were on a swim team. Ann and Neil played soccer. We encouraged each one to pursue individual interests and goals. When the children neared the teen years I began taking each child out to lunch, shopping, etc. by themselves about once a month. For a short time each one had my undivided attention.

I fear that I neglected our other children while spending so much time on Susan when they were little and needed me most. I think for the most part they did and do understand. They have enjoyed the few benefits of having a sister with FOP: an international circle of FOP friends, and multiple trips to Walt Disney World and Philadelphia. It’s hard to know what they were thinking when they were little. It does not seem to me in hindsight that they were often angry or resentful of Susan or us. They fought, loved, and played as most siblings do. We did have a firm rule that physical violence was not allowed (Susan included). But I let them argue, even yell at each other, so long as it did not get physical. Gerich and Ann have born the brunt of helping care for Susan. Neil as the youngest missed a lot of that. Ann continues to be a major help, even though she lives in another city. However, they have all grown up to be very independent, self-reliant people now. Perhaps that is a consequence of having had added responsibilities as children—I don’t know. I hope so. We did the best we could under very difficult circumstances, and we have survived. Gerich is now 30, Susan 25, Ann 23 and Neil 21. Thankfully the children remain close. Gerich, Ann, and Neil all live in the Raleigh/Chapel Hill area, about two hours from us. We talk, E-mail and visit often.

If I had to give advice to FOP parents, it would be make sure you spend quality time with each child and try to enjoy and nurture each one's special qualities.

About the author

Dorothy Kadala is 58 years old and has been married 37 years to husband Phil. She was born and raised in Anderson, SC. She is the fourth of five children and has four children, so she knows a lot about siblings! Her family has lived in Wilmington, NC for the past 18 years, but lived in The Netherlands for 10 years. They have also lived in Louisiana, Florida, Georgia, and South Carolina for short periods. Dorothy graduated from the University of Georgia, majoring in education and library science. She has a master's degree from the University of North Carolina at Wilmington in Language and Literacy. She has been an elementary school librarian off and on for about 33 years. Because she has taken a number of years off, having and caring for four children, she still has seven years to go before retirement. She loves to read, do puzzles, and walk, preferably on the beach. Her favorite place to be is her family's cottage on Colington Island in the outer banks of North Carolina. "It is Phil's and my sanctuary," she says.

19.

My sister and I

by Annie Kadala

While I was trying to decide what to say about being a sibling of someone with FOP I was a little baffled about what I could say that would help other people. My first concern is that my experiences are so specific to my place in my family and my relationship to my sister with FOP that I wouldn't be able to help many people.

My second concern was trying to separate how I feel about FOP and how I feel about my sister. I guess I'm not sure where the regular sibling issues end and the FOP issues begin. Finally, my sister and I are now adults and our relationship has changed a great deal since our childhood. How do I explain all of the different stages we have gone through and how FOP has affected those stages? I realized that I am not going to be able to completely separate Susan and FOP, I am not going to be able to make my experiences universal, and I am not going to be able to address everything that Susan and I have experienced. So, I just want the reader to appreciate that everyone's experiences are going to be different, and that I hope what I have to say will help at least one person in dealing with FOP as a sibling or parent.

There are four children in my family. I am the sibling closest in age to Susan, who has FOP. I am also the only other girl in our family. From my experience, and from observations, I believe that the closest sibling in age and of the same sex to the person with FOP helps more than other siblings with FOP related issues. For instance, I can take my sister to the bathroom and I can help my sister in the shower, but my brothers cannot and don't. They help less because they aren't able to help more. Further, since I am closest in age to Susan we were together in school a lot, so I was on the bus with her and at lunch with her. I helped in these situations because I was around, but my brothers didn't because they weren't there. I have felt like my situation in the family is unfair, but I think what has helped is that my mother and father never forced me to help my sister. It wasn't a chore I had to do. My help was appreciated and accepted but not expected or required. The expectations in our family are that we work as a team. Everyone is expected to pull their weight, so if we don't want to help take care of Susan, then we need to do something else around the house that will allow Mom to help Susan. Everyone has to understand that there are extra responsibilities in a family with FOP, and however you help in the household is enough as long as you are contributing.

As I said, I helped my sister a lot on a daily basis when we were younger. I did my sister's hair in the morning before school (because she liked how I did it better than anyone

else), I rode the wheelchair accessible bus with her, I helped her in the lunchroom, and I even helped with the other students in the Special Needs classroom. Now, when I am home visiting, I help my sister use the bathroom, I help my sister take showers, I help my sister dress, I drive my sister places, I make my sister's meals, and I help make my sister comfortable. As we have gotten older and FOP has progressed, Susan has needed more help in everyday life. It is a hard transition for everyone to understand when a person is no longer able to do something for themselves that they used to be able to do. All a person, sibling or parent can really do is make the best of the new situation and be creative in finding a solution.

For instance, recently Susan went to get a hair cut and color. Sitting in her wheelchair, she got her hair cut and dyed, but the challenge was leaning back into the sink to wash her hair. To solve the problem we backed the wheelchair up to the sink, and Mom and Dad lifted Susan and tilted her back into the sink while the hairdresser washed her hair. We were all laughing at how ridiculous we must have looked, and my mother made me take pictures so that when we explained the situation others would see how funny it was. My favorite picture is the one I took from above where Susan is wearing a cheesy grin and everyone is laughing. In the end, although FOP makes life harder, our creativity in dealing with FOP also just gives us an opportunity to laugh at ourselves.

About the author

Annie Kadala is 23 and lives in Chapel Hill, NC. She is in her second year of working towards a Masters in Library Science. She has a B.A. in English from the College of Charleston in South Carolina and hopes to work in a elementary or middle school library. She notes, "I have experience as Susan's sister and was also the nanny of a person with FOP. It was interesting to help a family who were new at dealing with FOP because I cannot remember a time that I didn't know what FOP was."

20.

Living with FOP:

When wanting is not enough

by Sharon Kantanie

Most people would have a hard time imagining life with fibrodysplasia ossificans progressiva, an extremely rare genetic disease in which bone grows in muscles and connective tissue, leaving joints of the body virtually frozen in position. Essentially, the body is forming an extra skeleton. Having lived with FOP for most of my thirty-eight years, I would have a hard time envisioning life without it. FOP has brought me great pain, many tears, and sometimes alienation from a world that seeks out perfection. FOP has also taught me some important lessons about myself and the world in which I live. I readily admit that at times my feelings about FOP depend a great deal on how closely I feel its presence as a silent enemy within my own body. Yet at all times I cannot help but think that the worst parts of my life are intricately tied to the best parts of my life—that my struggles have made the good things in my life count for more.

FOP can strike as a result of trauma as simple as a bump or fall. FOP also strikes for no apparent reason. There are also times when, in spite of obvious trauma, FOP does not appear. In other words, I never know when or where FOP will show up. Was it something I did, and something I could have prevented? My parents probably always will worry about such matters in an understandable effort to protect the child they created, and I can't blame them for that. But if given a chance to lead my life over again, I think I would still make the same choices, even those choices that hurt me in some way. I don't want to be so afraid of getting hurt that I forget to enjoy life.

Though it might sound strange, as a child I never thought of myself as different, even though there were certain physical things that I knew I was unable to do because I could not raise my arms. One particular incident stands out in my mind because it changed all of that. One day, when I was ten years old, my teacher saw me sitting on the sidewalk during recess while the other kids played a game called two-square. To play this game, two children bounced a ball back and forth in a grid until one failed to hit the ball. The teacher convinced me to try it and told the other students to “go easy” on me. This special instruction soon proved to be unwarranted, as I quickly turned into a very capable player and even moved to a four-player version of the game for a bigger challenge. It was the first time I had done anything physically challenging in my life, and it felt good.

My parents encouraged me and painted a court in the garage where children from the neighborhood would come over to play after school. Then, on the last day of school, I fell, injuring my knee. I did not give the incident much thought, for at that time I felt that it would take a much more traumatic event to bring on FOP. I soon learned how wrong I was. That incident started a painful four-year cycle in which I lost the motion in my left hip and knee. Since the age of twelve, I have relied on crutches to walk. I also lost the trophy of innocence, my sense of invincibility. I learned how simply things could change. But even if I could turn back time, I would still choose to have the experience despite the pain, both physical and emotional. I think that time means more because of what I have gone through since then. And, in the midst of the pain and tears, I was introduced to the person who remains my friend almost thirty years later. Our friendship is one of the few relationships in my life where I have never felt guilty about being needy, dependent and demanding because she chose to be my friend both because of and in spite of my disability. And I did not need her so much as we needed each other.

FOP affects each person differently. While there are certain similarities among people with FOP, there are as many differences. It is impossible to predict who will be affected minimally and who will be affected more seriously and more rapidly. There is one certainty about FOP: it is progressive. It also has a way of sneaking up on a person when least expected, at a time when the complacency of everyday life has set in—just at the moment when the latest limitations no longer seem the worst thing imaginable.

This begins a phase which I refer to as “When the wanting is not enough” because wanting the pain to stop and wanting life to return to the way it was is not enough to stop the pain, or to stop life from changing. But ultimately “When the wanting is not enough” is not about resigning yourself to the fact that you are not the one in control of your own body. It is about recognizing that things don’t always turn out exactly as you had planned. It is about fighting to change what you can and accepting the things that you can’t change so that you can go on living. It is not an experience that is reserved only for people who have FOP and other disabling diseases, but it is one that people with FOP must deal with earlier in their lives and on a more regular basis.

What is the hardest part about having FOP? Sometimes it is the pain. Sometimes it is that only one in two million people can understand what you are going through. Sometimes it is knowing that you are often dependent on someone else for the little things that most people take for granted. Small victories, like surprising my mom on Mother’s Day by dressing myself for the first time at the age of twelve, sometimes count for much. In general, however, I think that FOP is probably harder on parents and other family members than it is for the person who has FOP. I do not remember what it was like not to have FOP. So although there have been times in my life when I wished my condition would go away, I really cannot picture my life any other way. For me, I think the hardest part has always been not knowing what the final outcome of a flare-up would be and how much motion might be

lost. It is hard to make necessary changes to one's life until these facts are known. The interim period is one of fumbling in the dark and struggling to cope.

FOP controls my body, but it does not control my life. I know I am a stronger person because I have FOP. Dealing with FOP has given me the self-knowledge and strength of conviction that some people never find. We all have certain strengths and certain weaknesses and certain problems that we must overcome in our lives. Unfortunately for me, my weaknesses are the visible kind. They are the first thing people see and occasionally the only thing that people see. Fortunately, the people in my life, my family and my teachers, expected nothing but the best from me, and as a result, I have expected nothing but the best from myself

Rather than hindering what I can do in my life, sometimes, in what might be the ultimate irony, I think that I have accomplished more in my life because I have FOP. Or at least, I think my life has more meaning because of the path I have chosen, a path influenced by my own life experiences, good and bad. There are two accomplishments of which I am most proud. One is my career as teacher and tutor, though not because of any doubts on my part that I could do it. I figure that if I can get through FOP, I can get through anything. The other is my participation in the International FOP Association.

At times, my decision to become a teacher surprises me. All throughout college, until I applied to graduate school, I was determined not to be a teacher. Adolescents, often caught up in their own worlds, can be unwittingly cruel to a person with a disability. At least that was my own experience. To make matters worse, I was (and in many ways still am) painfully shy and afraid to reach out to people for fear of being rejected, or out of determination not to be a burden to other people. My high school years were at times bearable and at times miserable. I was determined to distance myself from those years. I also knew that teachers were overworked, underpaid, and sometimes underappreciated. In the face of these cold, hard facts, I have since decided that I did not choose teaching as much as it chose me.

As a teacher, I was pleasantly surprised at the open natures of many of the students I taught. If given the opportunity, they were not afraid to ask questions of me. "Why can't you sit down? Is FOP painful? How many people have FOP?" In all seriousness, a student once asked me whether I sleep standing up (as I am unable to sit). I easily answered questions about myself, questions I wish my classmates years ago had the courage to ask. I find that such questions served as a means to get past my disability. Undoubtedly, the hardest question a student ever asked me was "Did you like it as a student here?" during an occasion in which I went back to teach at my old high school.

I use my past experiences to make myself a better teacher and tutor. I try to have compassion and patience which my peers often lacked. I look for strengths in my students before I look for signs of weakness. I aspire to teach them that they can do anything they set their minds to do if they want it badly enough. I give them my best and expect nothing but their best in return.

The other thing in my life of which I am proud is the role that I played in the IFOPA. It all started rather inauspiciously. In 1989, I knew of no one else with FOP. Now I know of hundreds, many of whom I have met personally at IFOPA family gatherings. And when I was first diagnosed with FOP, my family was given very little information. As a result, one of the first things I wanted to do when I found the IFOPA was to ask if I could start a resource center for families. Little did I know that would later lead to writing and editing guidebooks and newsletters, publishing and managing a website, coordinating an E-mail discussion group, answering e-mails from all over the world and coordinating international meetings.

A discussion of the impact FOP has made on my life would not be complete without mention of the impact it has had on my entire family: my parents, a younger sister, her husband and their two wonderful children. I know it has been hard for them to watch me in pain and not be able to do anything about it. At times they have all made sacrifices for me. There were times when my sister did not get as much attention as she deserved because my needs were more pressing. There were times when my parents put aside their own needs for mine. My parents have often served as my arms and my legs, doing for me the things I could not do for myself. In college and graduate school, my parents drove me to more classes than I can count, and my mom logged many hours in the library searching for books I needed from stacks high and low. It is only because of them that I am where I am today. I sometimes feel extraordinarily guilty that I demand so much of their time. At other times, I am selfishly torn between their needs and my own. When I was diagnosed with FOP a little over 33 years ago, none of us knew what lay ahead. Thankfully, we took things one day at a time. I am not sure if you can take FOP any other way. I think we have all learned a lot in those years about what we could handle. Together, we have done our best to see a world full of challenges, rather than a world of problems, a world with possibilities, rather than a world of limitations.

About the author

Sharon Kantanie lives in Brentwood, Tennessee with her parents, and is fortunate to have her sister's family living nearby. She is 38 years old and was diagnosed with FOP at age six. She has a Master of Arts in Teaching from Vanderbilt University.

21.

Encouraging independence

Parenting children with FOP

by Sharon Kantanie

As an adult with FOP, I've often found myself saying to parents that I think that in some ways it is actually harder to be a parent than to have FOP. Parenting is difficult enough without the obstacles that FOP throws in the way.

As I thought hard about what to write that could possibly make parenting a child with FOP easier, I realized that there is an important lesson in the Disney-Pixar movie *Finding Nemo*. In the movie, a happy fish couple embarks on the adventure of parenthood until tragedy strikes, leaving dad Marlin to be a single parent to little Nemo, who is born with a deformed fin (or as Marlin tells Nemo a "lucky" fin). This sets the stage for Nemo's father to be overprotective and willing to do everything he can to keep Nemo safe from harm. That's understandable. However, this backfires as Nemo asserts his independence by rebelling against his father, and in doing so gets caught by a scuba diver and ends up in an aquarium. At this point *Finding Nemo* follows two stories. One is the tale of Nemo, who must use the skills he learns to find his own way home. The other story is a father's journey to find his son, which is in some ways just as much about the process of learning to let go as it is about the two fish making their way back to one another. Dory, a friendly fish actually states the lesson best. When dad tells Dory, "I promised him that I would never let anything happen to him," Dory responds by saying, "*That's* a funny thing to promise. Well, you can't *never* let anything happen to him—then *nothing* would ever happen to him. Not much fun for little Nemo." The challenge of parenting any child, and particularly a child with a disability, seems to be knowing when to be there for your child, when to let go, how to pass along the skills needed for your child to make his or her own way in the world, and how to have a little fun along the way.

One of the first things that FOP parents note is that it's important to create an environment where their children can do as much as possible. As RoJeanne Doege-Floyd, mother of Jasmin, age 13, states, "I try to encourage Jasmin to do everything that she is able to do on her own. The countertops in our kitchen are lower so that Jasmin is more comfortable getting her own snacks and food items. There's also a Lazy Susan.... We have moved all of her favorite foods to shelves within reach in the refrigerator." Some families have made other modifications to their lifestyles, such as living in one-story homes, adding

motion-controlled light switches, and making sure that floors are free from clutter. Tools like grabbers and dressing sticks can also make performing some tasks easier. (See Chapter 25, “Finding resources,” for more information and additional ideas). Chores are another good way to promote independence.

At times it can be difficult to encourage self-reliance while ensuring safety. As Carol Zapata-Whelan, mother of Vincent, age 19, notes, “This is a very hard balancing act, and it means preparing ahead of time to control the variables that one can control to ensure safety, telling others about FOP, taking necessary precautions, judging the risks versus merits of certain activities—and then allowing one’s child to have fun.” As Lori Danzer, mom of Erin, age 9, adds, “I can tell you that the panic never goes away, but it does get better. There will come a time when letting go is the only option. Believe me, it’s difficult, but you will build a trust that it’s going to be okay.” Lori recounts allowing Erin to go on the jungle gym at school. “She was so upset about not going on it with her friends. I finally talked to the school and worked it out with them and trusted that Erin would use her best judgment. That one decision gave the power to Erin, and it was important to her.” Debbie Hazlett, mother of Tim, age 11, expresses similar sentiments about letting her son do what he can. As Debbie says, “I want him to experience things while he can. If his immobility increases he may not, for example, be able to play in the snow... so I let him do this with his brothers even though I’m afraid he might fall.” Each family must weigh the options and decide which activities are worth the risk.

Irene Snijder raises a very important point. When physical limitations are present, it is just as important to encourage mental self-reliance. Irene’s daughter Tess is 16 and enjoys studying and preparing for university. Carol Zapata-Whelan’s son Vincent is in college and planning to go to medical school.

Inevitably there are going to be times where FOP limits certain activities. During times like this, parents note that they let their children express their frustration and help them find other opportunities. As Debbie Hazlett states,

I tell him that he may not be able to do certain things, but there are other opportunities that he has that other children do not, for example meeting a boy from Sweden (at a meeting of FOP families). Tim has also done Boy Scouts, which I didn’t allow his brothers to do because they were involved in too many sports.

Whenever Carol’s son Vincent gets discouraged, she notes that she tries to be there to provide understanding and encouragement. “I’ll say that I’m not happy about that either, but there are so many things that you can do. Let’s think and/or plan some.” Vincent was in the marching band in high school and continues to play in his university symphony. He’s

the family computer expert—something he seems to have in common with others who have FOP—and he’s been involved in numerous school clubs and projects.

One thing that you tend to hear from FOP parents is that children with FOP are generally very determined with a good sense of their own limitations. RoJeanne says, “When Jasmin feels safe and trusts her environment or the people she is with, she knows that she can step out. There have been numerous times when she has felt quite proud of herself for accomplishing what she perhaps didn’t think she could do.” That includes going on a two-night retreat with her confirmation class, as well as accompanying a sophomore at the high school to learn more about the experiences that will await her there. Irene Snijder has noticed that her daughter has gradually gained self-confidence and accepted FOP more as time has passed. For a while Tess had a boyfriend and was hesitant to tell him about FOP. When she did, everything was fine, though Tess broke off the relationship “because she wanted some freedom back.”

The clearest message that one receives from talking to parents of children with FOP is that they somehow find a way to look past the fear of FOP and focus on more positive aspects of life and being a parent. Lori Danzer, Erin’s mom, notes,

When Erin was young, I was constantly running my hands over her head and back, always looking for something. When she was about four, she said to me, “Mommy, I’m fine.” I realized that I was putting my fear on her. That is just no way to live. Now I let her tell me when something is wrong. It’s better for me and her. I try to put a positive spin on every situation for Erin. I believe it helps in building independence and happiness for any child . . . children need to be children. They have a life to live, and living in fear only takes that from them. It’s amazes me how children figure it out.

There is a story about parenting a child with a disability called “Welcome to Holland” by Emily Perl Kingsley. She compares the experience to being full of excitement after planning a trip to Italy and then suddenly finding out that the flight plan has been changed and you have arrived in Holland. At first, there is tremendous disappointment that the experience isn’t going to be what you had hoped. Then, if you are fortunate enough, you realize that Holland is . . .

just a different place. It’s slower-paced than Italy, less flashy than Italy. But after you’ve been there for a while and catch your breath, you look around . . . and you begin to notice that

Holland has windmills . . . and Holland has tulips. Holland even has Rembrandts.

Getting to the point where you can see the beauty in something that was as unplanned and unwanted in life as FOP is hard. It's something that takes time and more time, and none of us will ever be perfect at it. I truly admire the moms and dads who work hard to create a bright future for their children. I would also like to think that, as in *Finding Nemo*, parents teach children, and children sometimes teach parents. As Carol Zapata-Whelan states,

I think it is important to live every day fully, one day at a time, and not stress out over what could or couldn't happen. Often, our fears about our FOP children are unfounded. I think they develop an internal strength and acceptance of life as it is, a will to overcome, and a capacity to adapt to the unexpected that makes them unique and amazing. We just need to offer a steady, encouraging presence, take wise precautions, and have faith that they will make their way.

About the author

Sharon Kantanie lives in Brentwood, Tennessee with her parents, and is fortunate to have her sister's family living nearby. She is 38 years old and was diagnosed with FOP at age six. She has a Master of Arts in Teaching from Vanderbilt University.

22.

Redefining independence

Adults with FOP

By Myra Bellin

Knees and shoulders and hips are made of bones and ligaments and muscles and cartilage, all calibrated for movement. But with FOP, extra bone growth hampers movement and, as the disease progresses, joints which no longer function properly make it difficult to sit or walk or bend or reach. These difficulties affect many other aspects of life. Activities such as showering, eating, and dressing may be impacted, which means that issues of independence accompany the condition. Can people with FOP live alone? Should those with FOP rely on family or hire people to help them? Where do they find caretakers? Is it possible to help family members with FOP maintain independence? How?

Adults with FOP have many different lifestyles. Steve Eichner is 37, married, has a PhD, and now works as an IT Program coordinator in Texas. Sharon Kantanie, age 38 spent a semester away at college but returned home after a flare-up locked her hips, earned a masters degree in education, and has worked as both a teacher and a private tutor. Roger zum Felde is a German man in his forties who lives in an apartment near his parents. He worked in the chemical industry for fifteen years, and then continued to do accounting work in a home office for five years. A guest appearance on a TV talk show about FOP opened the world of journalism to Mr. zum Felde, and he now helps write and produce documentaries about FOP. Tonya Barnes, 38, lived first with a boyfriend and then on her own for several years, but moved back home to help her mother after the death of her father.

Because of the physical limitations imposed by FOP, the condition often imposes the necessity for physical help from other people. Obviously, the amount of help needed depends on the level of disability. Steve Eichner finds it difficult to reach for items when he goes supermarket shopping and says that most people are quite willing to help if he just asks. Steve has not needed any additional help with his daily life, but Sharon, Tonya and Roger all have hired caretakers. Each has arranged for hired help to suit their own needs and schedules. Roger has arranged for different people to help him with different things, including dressing, food preparation, or massage. These hired caretakers help him retain his own apartment within a residence for older adults.

Sharon and Tonya also have hired caretakers either on a daily or hourly basis to assist them. Tonya finds there is a real benefit to hiring people because it “lets you stay independent from your family. Make your own decisions.” The disadvantages, she feels, come from the need for relinquishing privacy—outsiders learn the personal details of her life. Tonya emphasizes that it has been important to be very clear and strong about her needs with caretakers, instructing them about how to help her so they do not inadvertently cause injuries. Trust is an important issue with caretakers, and she says it is important to listen to your own instincts about people and to speak up about matters that cause concern. Tonya says that interviewing and hiring caretakers, while a difficult process, has helped her take control of her life

Sharon, too, likes having caretakers because they not only help her, but they give her parents more freedom with the peace of mind that comes from knowing that she is cared for. Family doctor referrals, answering or placing newspaper ads, word of mouth—all are methods for finding people to help.

Adaptation is an important word for those who live with FOP; in one way or another, those with FOP alter their living environments to enable them to function as independently and effectively as possible. Tonya Barnes offers some examples of how she adapted her environment.

I had a ramp installed when I could no longer climb steps. One doesn't realize how restricted your life can become when you can no longer climb stairs. I had a walk in shower installed, purchased a fully electric hospital bed, a recliner lift chair and my stand up wheelchair. When I could no longer get myself in or out of bed, my caregiver hours increased.

When Sharon Kantanie left home for college, the months she spent living on her own were aided by making sure the environment could be adapted to her needs.

We found adaptive tools so that I could brush and wash my hair, dress myself, etc. I learned to drive. We found a college that was close enough to home but far enough away that I would feel “on my own.” We made special arrangements at the university.

One reason that Steve Eichner is happy with a recent move to Texas is because a house with only one floor is both more feasible and more affordable there than in his prior location near Washington, D.C. And Roger zum Felde is constantly rethinking and revising

his environment to maintain his independence—for example, he now has a bed he can enter standing up which then lowers electronically.

Adults with FOP may need varying degrees of physical help, but, as Tonya Barnes put it “our bodies may be restricted, but our minds work great.” Maintaining and fostering a sense of independence that is separate from physical needs is a priority and family support is crucial for this, particularly in the early years. Steve Eichner sees it this way:

Even though FOP has certain challenges, you can't let it stop you and your child from experiencing life. You need to create and foster independence early. While there are a vast number of technology solutions (reachers, special tools, etc.), instill creativity and problem solving. It's amazing what one can do with a coat hanger and some determination.

Tonya Barnes expresses similar sentiments and cautions parents not to keep a child with FOP in a “bubble” –rather; they should encourage their children to experience life.

It is very important to let your child be a child! Let them know the boundaries and trust that they will make good judgment. I am thankful that my parents didn't restrict me. I treasure my childhood memories and all that I was able to do. I rode a bike, played soccer, hide and seek, and drove a car for 2 years.

Maintaining an active stance in the face of FOP has helped those with the condition in all areas of independence. Steve Eichner is still able to sit because of a medical procedure in his teens. Recognizing that his left hip was likely to fuse in a way that would prevent him from sitting, his doctors attempted to trump the process by putting him in a body cast when he was thirteen—the thought was to control the placement of the fusion. Because of this stint in a body cast, Steve is now able to sit and can use an electric wheelchair, which he often does to put others at ease. “A wheelchair does not challenge people's norms,” he says. And he feels that others are more comfortable seeing him in a wheelchair than seeing a posture and gait, which seems, to them, awkward and strange.

Roger zum Felde is constantly rethinking and revising himself in face of the encroachment of FOP on his mobility. When it was no longer possible for him to use his left hand to write, he trained himself to work with his right hand. It was always important to Roger that he earn his own money and he had a job for many years. When additional physical restrictions made going to a workplace on a regular basis too difficult, he maintained an office at home for five years and when that too became difficult, Roger diverted his energies to working on journalistic pieces about FOP.

Sharon Kantanie feels that it is important for her to focus on those things that she can do. “Everyone has strengths and weaknesses. Mine are just the physical kind, and that leaves me dependent on other people for those things.” She still does some tutoring and spends a lot of time at the computer managing and editing writing projects, keeping up correspondences, and editing home movies. Tonya Barnes ignored the advice of a high school counselor and studied accounting. She has no regrets. She was able to work as an accountant when she graduated high school, lived with a boyfriend for several years and then lived alone. Although she no longer has a job, Tonya is glad to have mastered marketable skills. She loves quilting now, and gets help occasionally in pursuing this hobby since she can only reach with one arm.

As far as advice for those with FOP and their families, Roger zum Felde emphasizes that it is important to be strong. His family helped him by never saying that he couldn’t do something, rather they worked hard to try to help him accomplish what he wanted whether it was going to school with his best friend as a child, helping redesign the bathroom or fighting for the financial benefits due him under various governmental regulations and programs.

Sharon Kantanie acknowledges that “It’s not an easy thing, even if you’ve been dealing with it all of your life. But I think overall it’s better to focus on the positive rather than the negative, the things you can do rather than the things you can’t.”

And Steve Eichner has the following advice for families:

Just do it! You can’t let fear of what could happen with FOP immobilize you—if you do, the disease “wins” outright. As one looks at career paths (always an uncertainty), try to evolve yourself into a world that doesn’t require as much travel or physical impact or plan, through education, to have a migratory path that leverages your physical experience if you become more limited in movement later on.

Careful planning helps, as does a little luck.

About the author

Myra Bellin is a freelance writer who lives and works in Philadelphia, PA. Her interest in FOP began after a visit to the Mutter Museum when she began to wonder about the life that once animated Harry Eastlack, a question which led her to meet members of the FOP community. She has published in The Rambler, The Philadelphia Inquirer, and Ceramics Monthly.

23.

FOP and school

An overview and discussion

by Susan Duberstein

“It’s the first day of school.”

Those words conjure up images and emotions for just about everyone. Kids may think of the smell of new erasers, the excitement of getting to ride the school bus for the first time, shyness at meeting a new teacher and classmates. Teachers must all know the anticipation of what a new year and a new class will bring, as well as the joy of seeing returning students, now moved on to the next grade, who will wave to their old teacher on the way down the hall. Or—for parents—the thrill of watching one’s own child set out on that same great adventure, pride in the accomplishment, and just a little trepidation at the thought of letting them go. (And maybe the excitement of looking forward to a few hours of free time!)

I revisited that thrill beginning in August of 2001, when I started back to school to begin the pre-requisites for medical school. As a “non-traditional” student who had been out of the classroom for more than eleven years, I was excited, enthusiastic, and scared to death.

Every new experience brings new challenges, and every new challenge has its potential difficulties. For parents of children with FOP, or indeed with any chronic medical condition, these concerns are more concrete. All parents worry about their childrens’ safety, their acceptance by peers, and understanding on the part of teachers and school staff; these issues are just more specific and immediate to FOP parents.

The publications listed in the IFOPA’s *Catalog of FOP Resources*, available at the IFOPA website at www.ifopa.org, particularly the “FOP & School” article found in this collection, do a wonderful job of delineating the sorts of questions that should be asked, the resources that should be explored, and the possible solutions to technical or physical obstacles in the classroom. Additionally, the FOP newsletters and the FOPonline E-mail newsgroup are useful ways of passing on the good ideas others have come up with and the methods others have devised to successfully deal with the unusual and individual difficulties that may be faced by children with FOP when entering the school years.

This article is not intended to reinvent the wheel, but to provide a more personal overview of advice from both parents and educators who have had experience with FOP children. I also spoke with a number of pediatricians with expertise in children with special medical needs.

I'm a future pediatric specialist, and one of the catchphrases of our trade is "anticipatory guidance." They teach us that our job is not only to examine the child and to treat any problems that brought them into the office, but to think ahead and bring up issues that may not have yet occurred, and to ask for the answers that a parent may not realize are important or think to provide. I would submit that this is exactly the same outlook a parent of a child with FOP needs—to be prepared with not only the kinds of questions that should be asked, but also the kinds of answers that may be provided from both sides of the fence. In the course of my research, it seemed like the more questions I asked, the more I thought of to ask, and that's probably the main take-home message for everyone: keep the lines of communication open at all times because you never know when someone will come up with something you wish you'd thought of earlier.

The students, parents, and educators who contributed their thoughts will not be identified by name, both for reasons of privacy and because I didn't want the discussion to be specific to any one location or situation. Some issues were purely technical, some were quite personal, and some were as applicable to any child in the world as to one with FOP. Overall, it was clear that there were a number of constant concerns.

Safety

Clearly, safety was far and away the foremost issue for just about everyone I spoke with. With the very real potential for permanent consequences to seemingly minor physical incidents, safety has to be paramount. Since it's unlikely that anyone who works at the school will have heard of FOP before, the family also has to instruct, and this can be a daunting task.

The heartening thing I found was that the vast majority of stories from both educators and parents were overwhelmingly positive. The first thing one teacher told me was that "it was extremely important that [the classroom] was a physically comfortable and safe place to be." She went on to add, "Our administrator held a special meeting with [the student's] physician to help key the entire staff in on the importance of taking preventative measures," she added, and this is a great idea. Bringing participants from different spheres of a student's care together is a terrific way to make sure everyone has the necessary information. (I think it's important to add that privacy issues are very often a serious concern for parents—as a pediatric specialist, I consider that a very special trust. Medical privacy laws are stringent for

a very good reason; no one needs to have access to your child's medical records who is not directly involved with treatment. However, education about FOP itself is not specific to any one child and education is key to easing fears and concerns about caring for a child with FOP. It is not necessary for non-medical personnel to know details of a child's treatment, but is essential to understand the reasons behind the need for safety and protective measures.)

Going through the daily routine with the teacher was brought up a number of times as an example of a good way to be sure there are no unforeseen difficulties; this is usually done in the process of preparing an IEP (Individualized Education Program) or other documentation. Make sure to review every part of the day, and ask to see the schedule on paper. "You can think you've gone over every possible problem in the classroom and then realize that there are two steps to get down to the cafeteria," one mother mentioned ruefully. "And then they suggested that [the child] just eat lunch in the classroom!"

On that note, it's an unfortunate reality that liability is often brought up as a reason to limit the child's activity, rather than as a reason to find a solution that is inclusive. School administrators may be overly concerned with potential dangers, and teachers or aides may be afraid of not being able to provide adequate care for the child with special needs. At least one parent mentioned having to hire a lawyer to counter a school's fears of liability, and this is regrettable. But the bottom line is that it is the school that should accommodate the student, not the other way around. "That [they] had to work to make things safe...was just the way it had to be," another parent stated flatly. "Because we would not accept being left out." Many of these concerns can be addressed with sufficient information and by working together to create a clear plan both for day-to-day issues and for steps to take in any potential emergency.

You are of course your child's primary safety officer, but administrators, classroom staff, and other students alike must grasp the necessity of taking precautions. "It's not personal. I don't trust anyone" is one mother's mantra, and everyone does indeed have to be on the same page on this topic. Additionally, the child should also be supported and encouraged to be proactive in his or her own care, as appropriate to the child's age. He or she is the one person who's always guaranteed to be present in every possible situation, after all!

Understanding

One of the great things about pediatrics is that kids, no matter what their medical problems, are still kids. It's a huge part of what I love about my future profession. "I wanted to feel like as normal a kid as possible without too many things separating me out as different," one

FOP adult recalled. This was echoed many times as parents, former students and educators alike related story after story of ways they found to fit in, to participate, and to enjoy their school experiences along with their classmates.

Peer relationships are key to these experiences. Other kids will not know what FOP is any more than adults will, but they too can be taught. One particularly self-motivated young man actually ran an assembly for his entire grade level at numerous points in his scholastic career to introduce his classmates to FOP! This level of participation and independence is extraordinary, and some children may prefer to work with smaller groups of friends, but recruiting the support of the peer group is indispensable.

Classmates can help a fellow student in ways that no adult ever can—they are the ones who share class time, who play the same games, laugh at the same jokes, and make fun of the same dumb things that grown-ups say. Student helpers can assist with simple tasks like carrying books or lunch trays and “tying gym shoes” as a mother mentioned wryly.

More than one parent spoke with me about the kinds of life skills that aren’t taught in the classroom: assertiveness, empowerment, independence, socialization. “[The playground] is where all kids develop the ‘real’ skills they need to deal with life post-school,” one parent wrote. Participating in as full a range of activities in as ordinary a way as possible is to be encouraged; having the support of friends in the classroom can smooth over many minor difficulties. “We also encourage him to be a giver as well as a taker and to help other students out,” that parent continued. Reciprocity is a big part of every good relationship—if another student helps carry your books, maybe you can help them with their math homework.

Older “buddies” from upper grades can also be hugely helpful as “guardian angels,” as one mother described them. During my training, I often noticed how unusually mature many kids with medical issues can be, and having a more mature “big brother/sister” to talk to can work out beautifully. Of course, a student’s own siblings, if there are any, are a big part of socialization too—but maybe not quite as cool as the Big Sister in the eighth grade when you’re only ten.

Balance

The take-home message here: nothing is more important than the child getting a good education and feeling comfortable, accepted and actively involved at school. Second to safety issues, this idea probably came up the most. The realities of the need to protect children physically can often be at odds with the desire for them to have as normal a school

experience as possible, and this can be one of the most difficult parts of FOP for both parents and children.

Yes, it's inescapable that FOP kids are "different," but not in most ways. FOP may be a complicating factor, but complications can be gotten around. Fortunately, as was abundantly clear, creativity is practically unlimited where these kinds of solutions are concerned. Activities can be adapted, provisions for extra help if needed provided, and emphasis placed on pursuits in which the child is not limited.

One parent who was having some issues with inclusion at her child's school was very eloquent in her frustration: "I feel very strongly that with [the school] that the pendulum always swings in the direction of limiting experiences, and I'm always fighting that," she told me. "There are good reasons for staying, but I keep feeling like I'm on a tightrope between telling them why they have to be careful and why they don't."

Another, perhaps inevitable complication: as children mature, their desire for independence can come into conflict with "sensible" decisions. During my adolescent medicine rotation, a young woman I spoke with about her own chronic condition—not FOP—told me firmly that wanting to be "just like everyone else" was at the heart of a great deal of teenage rebellious behavior that led her to fight a lot of medical treatment and precautions. "I always felt like they were telling me what I couldn't do," she told me. "And I would be like, well, I'll find a way to do it anyway, so work with me and not against me, know what I mean?" Yes, ma'am, point taken.

The simplest and most heartfelt statement I heard on this subject came from a teacher: "Based on my experience, the advice I would give...is to interact with your FOP child in the same fashion that you would your other children. Respect them, love them and keep your expectations high."

Well said.

Resources

The previously-cited articles accessible on the FOP website go far beyond the scope of this article in listing all sorts of high- and low-tech solutions to common obstacles, so I won't go into listing all of those things here. Additionally, what resources are available to accommodate students varies so much by area and jurisdiction that it's impossible to make any generalizations, but the main thing to remember is that you may never know what may be available unless you ask. So ask!

Under this heading I'd mention the topic of personal aides. This subject is a matter of some delicacy, and there is no one right answer for every child. Some families have found that an aide is an indispensable resource for their child, and have forged incredibly close bonds with the special people who have worked by the child's side for years. For some, aides are seen as adjuncts that may be useful in certain situations where extra assistance is required, and for others whose children may prefer more independence, they are seen as unnecessary or even intrusive. Most school systems have provisions in place for personal aides; some may be individual to that child, or they may be responsible for more than one child with special needs. In any case, it cannot hurt to inquire what is available.

Some children do very well with minimal day-to-day help being given by their peers; some may need more individualized attention, and of course the question can always be revisited as the student's physical needs change and their level of independence and sophistication changes. It is of course important to consider the level of assistance that can be expected. School friends can help carry books or share notes, but would not be able to assist with more personal needs. One advantage of a professional aide is also that they provide a stable source of assistance where a teacher may be distracted or a student helper may be absent.

In deciding whether or not an aide is right for your child, find out what is available, remember that it is often easier to cut back than to decide midstream that more help is needed, and be prepared to adjust as the child's needs change in either direction.

As well as school aides and student helpers, resources include assistive technology. This ranges from accessibility for wheelchairs to things as simple as a tilted writing board to make note-taking easier, or a flag to hold up for a child who isn't able to raise a hand. Issues of mobility when changing classrooms might be solved by simply leaving class a couple of minutes early; several people mentioned having extra sets of books at home or in different classrooms so that they don't need to be carried throughout the day or using lecterns or needing a particular placement in the classroom to make participation simpler. Those are all simple things that can be accommodated in any classroom or school. Professional assistive technology experts do exist, although you may have to ask about them, and these specialists can be incredible resources in themselves.

In terms of finding funding and services, a number of people made the point that it is not enough to go by what any one organization suggests. The school may have a list, but so may your pediatrician, your church or other place of worship, government agencies and community groups. One parent noted, "The more people I talked to, the more people had the chance to help." The success of so many of the FOP fund-raising events speaks strongly

to this—people are willing to help but may not know what is needed or that help is needed at all.

Private versus public schools came up often. Some parents and former students talked about the smaller classes and more individual attention being positive factors in choosing private schools, while others mentioned the desire to have the child attend the same school as siblings or neighborhood friends as being significant factors in their choice to stay in the public school system.

Any child with a chronic medical issue will need some accommodations, even if it is just that they may miss school more often. Some parents find that private schools are more open to the kinds of adjustments that their child needs, or that access to occupational therapy or other extras was easier in the public system. The presence or absence of a school nurse was also mentioned in relation to this decision. Another good point was that in most areas, a child's enrollment in a private school does not mean they are not entitled to public resources.

Advocacy

This category too came up time and time again, and the advice was loud, clear and unequivocal: advocate early and often. It's never too early to begin discussions, and important to keep careful records of who has been spoken with, what topics have been discussed, and what expectations are for the future.

Be prepared! “Always go with a list of questions and concerns as to how they will help your child create an environment where he/she can learn the way they need to,” was one mom's very wise advice. The suggestion to write things down cannot be overemphasized. Too often, as different subjects come up and discussions go in various directions, important points are forgotten. For formal meetings, especially if there are going to be multiple people involved, e-mailing or faxing a copy of the topics to the participants beforehand can also be helpful. Not only does a rough agenda allow everyone to be sure all the issues are covered, but if there is any information that needs to be researched beforehand, it will give administrators, teachers or others the chance to prepare that. Nothing is more frustrating than finally gathering all the caregivers in one room, only to hear over and over, “That's a good idea—I'll have to look into that and get back to you.” Maximize the time you have together with the caregivers in your child's life by being organized beforehand.

Some families have social workers or other professional advocates whose job it is to help smooth the processes. There are times when expert assistance is invaluable—it makes no sense to spend hours poring over instructions if there is someone around whose job it is to

fill out the forms! Whether or not you have that option, having a close friend or family member to be a “second set of ears” can also be a great help. “It keeps the school honest, if you will,” I was told by an FOP adult. “And erases the possibility of ‘he said/she said’ after the fact.” Great advice.

You are your child’s first and primary advocate, without doubt. It sounds like such a cliché, but it’s nonetheless true. No one else is ever going to be as intimately aware of your child’s needs—with one exception, and that is the child him- or herself. At age-appropriate levels, again, it is also important that the child also begin to be involved with their own school planning and needs. FOP kids go through the same stages of normal development as every other child, and when one mother told me “[My child] is very much his own best advocate,” that statement resonated with me very strongly as a declaration that this was a child who was ready to take on the world. What more could you ask?

It was a pleasure and a privilege to be asked to contribute to the new Guidebook. The part of my future career that I look forward to the most is the opportunity to be involved with both children and parents throughout their lives, and it has always been a joy to be honored with their trust and their confidence. Thanks to everyone who responded and for all the inspiring stories and suggestions.

About the author

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24.

FOP and school

Ideas and resources

(Adapted from the article titled “FOP and School,” found in the IFOPA’s Catalog of FOP Resources, available at the IFOPA website at www.ifopa.org)

FOP does not usually affect the person’s intelligence or cognitive abilities. It does, however, negatively affect the child’s ability to safely access the school premises, use and manipulate the school’s materials, and function academically in a typical way. The needs of a child with FOP in a school setting have to do with gentle handling, preventing falls or bumps; and positioning the child so he/she can see, hear, and gain access to places (desk, floor) and school materials with the least amount of strain on his/her body as possible.

The various issues that arise during school can be addressed by age level: pre-school and kindergarten, elementary School (1st to 5th grades), middle or secondary school (6th to 12th grades) and transition, and post secondary education & employment. As the child passes from one setting to another, many of the adaptations stay the same, some form the basis for new accommodations, and some are no longer necessary. Note: This article refers to United States laws only (see end of this section for description of these laws), but the adaptive suggestions made in it are universal.

Pre-school, kindergarten, and elementary school

During the toddler years, children with FOP are mainly like their peers. Some may have stiffness in their upper bodies and balance problems, but in general their bodies function like their friends. This means that the primary issue at this time is the overriding need to prevent trauma that may start a flare-up. When preparing for pre-kindergarten or kindergarten for a child with FOP, most parents have found that the best way to be sure everyone understands all the concerns is with a face-to-face meeting where the best interests of the child is clearly the focus. Such a meeting should include everyone involved with the educational experience of the child, including the new teachers, previous teachers, aides (if any), school nurse, and any service providers, such as occupational therapists (OTs), physical therapists (PTs), speech & language specialists, etc.

The decisions reached during this meeting should be written down to make it easy to bring other participants up to date and to encourage compliance. It can be done informally, or the plan can be part of a formal “Individualized Education Plan (IEP)” pursuant to the Individual with Disability Education Act (IDEA) or part of a written “504 Plan” (pursuant to the Rehabilitation Act of 1973). See below for more details. This meeting should take place before school starts to assure that the child’s inclusion will be seamless and natural. In general, the more you do before school starts, the better it will be for the child.

Concerns that may arise in pre-school, kindergarten and elementary school are listed below: *One-to-One Aide:* The parents must decide whether to request a one-to-one aide for the child with FOP. The aide’s primary focus during preschool and kindergarten would be to try to keep the child from sustaining trauma by watching for hazards of all kinds, such as slippery surfaces, tripping dangers, playground dangers, and other young children (often with poor impulse control). As children get older, the aide not only looks for hazards, but also assists the child in the lunchroom and bathroom (if needed), carries books, puts materials in front of the child (if needed), copies assignments, transcribes other work when appropriate to limit arm fatigue, and assures safety during transitions around the school. Whether an aide is the best way to keep the child safe will depend on the size of the class, the facilities and layout of the school, and the child’s degree of FOP stiffness.

Tables and desks: Tables and desks should be at the right height for minimizing the strain on the child’s neck and back while writing, drawing, and playing. To facilitate early writing skills, a slanted desk top or writing board (which can be made by taping two binders together) can help a child with restricted neck movement learn to write with less strain. A desk that can be adjusted to different heights and/or angles will be able to accommodate the varying degrees of back flexibility and arm motion exhibited by a child with FOP over the course of a year or several years. A “wheelchair cut out” design (which is curved to allow a wheelchair to get very close to the desk) can give the child with FOP the maximum space he/she can access with limited arm movement. An L-shaped, two desk set-up can also maximize the amount of space accessible to the child with FOP.

Paper and book holders: To assist with reading or copying, a page holder can be extremely helpful. For a child who cannot tilt his or her head down to look at a book resting on the tabletop, a book holder is essential.

Seating: The chair of a child with FOP must be very stable, possibly padded, and at the right height so that the child’s feet are always firmly planted on the floor for proper back alignment. Depending on the position of the child’s elbows, the chair may need to be armless. The chair may need to swivel.

Floor sitting: Circle or rug time may be difficult if the child can't sit on the floor unsupported. A stand alone booster seat, floor chair, big pillow or bean bag may help the child stay at the same level as his or her peers.

Foot rest: A foot rest can help the child maintain proper leg to hip position and thus reduce the strain on his or her back and spine while sitting.

Priority seating: Since students with FOP often cannot turn their heads due to neck stiffness, they should be seated where they can see the teacher easily and without strain. Also, seating the student in the front of the classroom can also accommodate the moderate hearing loss often associated with FOP.

Velcro holders for materials: School materials, such as pens, papers, etc. can be kept within reach of the child with FOP by holders fastened by Velcro.

Backpacks: To reduce the strain on the child's back, a rolling backpack can be very helpful, as can having a second set of textbooks at home and/or an aide to carry things.

"Pick me stick:" As a substitute for raising one's hand, many parents create a "pick me stick," featuring an item on a stick that can be waved to get a teacher's attention.

Test and work modifications: The child with FOP may need extra time for tests and assignments that require a lot of writing.

Cubbies: To foster independence, the coat hook of a child with FOP can be lowered to a height that the child can manage on his or her own. To reduce the dangers of crowds, the cubby itself ideally should be in an area less prone to the traffic of children all going to their cubbies at the same time.

OT and PT evaluations: Occupational therapy (OT) and Physical therapy (PT) evaluations of the child's hand and arm strength, fine motor skills, balance, ability to transition from floor to feet, and ability to toilet independently can be very helpful. As needed, the therapists can work on these skills, maintain current muscle strength and flexibility, and compensate for increasing stiffness as long as they appreciate how it needs to be done without straining the child's body.

Assistive Technology (AT) evaluation: Assistive Technology (AT) evaluation: Assistive technology is any item that helps a person with a disability perform a task. It includes low-tech items, like pencil grips, and high-tech items, like special computer keyboards. The

proper time for an AT evaluation is determined by the child's physical limitations and educational difficulties, and by the recommendations of the child's teachers, parents, and therapists. A good AT evaluator will be able to suggest time- and energy-saving electronic devices and software to help the child with FOP keep up with peers and conserve energy and arm strength. Some examples of AT are computers with accessibility software and hardware (trackball mouse, on-screen keyboard, word prediction software, Sticky Keys to make capital letters with one keystroke, speech recognition software, and handheld computers such as Palm Pilot or iPAC).

Independence: The school can assist in the creation of activities that will allow the child with FOP to develop his or her growing sense of independence and self-confidence, especially at school. This may include carrying messages to other teachers during times when the halls are quiet, helping younger children at lunch, policing an area for litter, etc.

Inspection: The parent can request to inspect the classroom for safety hazards, such as narrow or cluttered passageways, books on the floor, area rugs (which may need to be taped down), etc. both at the beginning of the school year and periodically throughout the year.

Physical education: During this period, the activities in PE will vary from simple safe activities, like games and exercises, to more active and/or risky activities, like gymnastics and soccer. These activities may be modified on an as needed basis for the child with FOP. The student might be excused from physical education, attend adaptive PE (a program created just for him or her), or receive time with a physical therapist. One school district arranged for the child with FOP to get her PE credits in a special swim class that helped her physical development in many ways.

Recess and playground: For a child with FOP, the safety factor is the biggest issue. If there is a one-to-one aide, he or she should stay close to the child with FOP and try to watch the area around the child for any dangers, such as tripping hazards, running children, etc. It is important to try to find ways for the child with FOP to be part of the child-initiated group activities despite the presence of an adult. This gets harder as kids get older because they begin to make up their own games and most of them involve running, climbing, chasing, wrestling and other things the child with FOP might not be able to do safely. To help involve other children in what the child with FOP is doing, the parent or teacher can bring in special materials or equipment that everyone can use during recess, such as sand toys, long-handled shovels for sand boxes, large pieces of chalk to be used for coloring or tick tack toe on a wall or ground, soft balls for kicking, catch, etc. The idea is to create games that will be fun, safe, and engaging.

After school activities: The child with FOP should be able to attend appropriate after school activities if they are being offered to the rest of the students. The school district may have to pay for the aide to attend these activities as well. This may depend on the child's educational classification and on the level of advocacy by the parent.

Transportation: If the child with FOP will be taking a bus, the bus aide should be educated about FOP, understand the need for safety and seat belts, and be given an opportunity to ask questions. Some children who have FOP receive door-to-door transportation, as opposed to being picked up at a bus stop, which would involve the risks of a walk and wait.

Safety rules: It is important that the other children in the class be aware of the special FOP related safety rules. Either the teacher or the parent may speak to the children in age appropriate language, telling them a little about FOP and reminding them to always push chairs in close to the table, hang up coats so that no one trips on them, not to push on the way to the door, etc. Parents should also discuss with the school how to care for the child with FOP during fire drills, real fires, and other emergencies, such as choking incidents.

Staff meetings: To spread correct information about FOP and the child's needs, the parents might meet with not only the child's teachers and therapists, but also ancillary school personnel, such as lunchroom staff, maintenance people, & other teachers.

"Sensitivity meeting:" Educating everyone about FOP—students, parents, everyone—may minimize teasing and bullying later. A sensitivity meeting can be as simple as a meeting of classmates and staff, a general assembly, or anything in between. It can be a real opportunity for educating the school about FOP and the issues of living with a chronic illness.

Communication: Some parents have written letters to the parents of all the children in the same grade as the child with FOP explaining FOP and how it is affecting their child. This is one way to be sure that other parents have accurate information about FOP, and not just second-hand stories from the kids. Since you opened the door to communication with a letter, it can make it much easier for other parents to talk with you about your child & FOP.

Emergency information: The IFOPA's FOP emergency cards should be kept in the classroom along with additional contact information of those to call in case of an emergency, such as mom, dad, pediatrician, etc.

Secondary school (6th to 12th grades) and transition

Reading, writing, researching, thinking, and extra-curricular activities all come into the picture. To help the student with FOP succeed, the previous adaptations should be re-evaluated to see if they are the best and most up-to-date options available. There may also be a few new concerns to be considered, such as the transition to the post-high school life of college and/or employment.

Personal aide: For a student in this age range, the aide's duties expand. She or he may assist the student with lunch and toileting (if needed), carrying books between classrooms, setting up materials (test tubes in science lab, etc), taking notes and copying assignments, transcribing other work when appropriate, and helping the student with transitions around the school so that they are safe and timely. The child and aide may need to leave each class five minutes early to avoid crowds of students in the hall.

Assistive Technology evaluation: At the same time that school work increases dramatically, FOP may be limiting the student's mobility. AT may now include mobility equipment such as wheelchair, scooter and cane, and more electronic technology, such as a laptop computer, accessibility software and hardware, a trackball mouse that is held in the hand rather than rolled on a desk, on-screen keyboard, word prediction software, Sticky Keys to make capital letters with one keystroke, and speech recognition software. Handheld computers such as Palm Pilot or iPAC are also useful for students with limited range of motion. In later years, another in-depth AT evaluation should be done to see if the AT should be updated again. For example, a desktop computer can be adapted to the current needs of the student with a cordless mini-keyboard, trackball mouse, Kurzweil software that allows the student to scan a book, read it and extract notes on the monitor screen, and a printer/scanner for the home.

Test and work modifications: There may now be a need for further test and assignment modifications, such as oral test-taking, extra time, less homework, submitting work written on the Palm Pilot (which means it would have to be brought home, downloaded, printed & returned to the school for submission).

SAT accommodations: The Scholastic Aptitude Test (SAT) offered by Collegeboard.com is a standardized test taken by high school juniors and seniors. The score is submitted to colleges as a way to compare students in the graduating class nationwide. The Preliminary Scholastic Aptitude Test (PSAT) is a "practice" SAT taken in the fall of junior year. PSAT scores are used to nominate National Merit Scholars.

Accommodations for the PSAT and SAT can be made for students with a disability. The College Board grants the accommodation. The high school counselor is the person to negotiate accommodations for you with the College Board.

We are aware of the following accommodations being provided to students who have FOP: extended time, such as “time and a half,” which is 50% more than the time usually allowed to students for the test (for example, 90 minutes for a 60 minutes test); a person to record/transcribe answers; being allowed to move around during the test when needed to release physical stiffness; alternative test location to facilitate access or special seating, such as taking the SAT in the counselor’s office; and divided seating, to accommodate a student who cannot tolerate taking the SAT in a single 4½ hour session, which is the standard time and a half accommodation for a 3 hour test, and therefore he or she takes the divided test in two sessions, 2¼ hours each, preferably on two consecutive days.

Transition: Public schools must address the transition to post-school life for students who have an Individual Educational Plan (IEP or IPE). Transition services begin when the student is age 14. The student and his/her family decide on the student’s goals for the future. School and community personnel work with the student and parents during the last years of public school to prepare the student for life after graduation. Vocational Rehabilitation becomes involved with transition services during the last year of high school. The transition services required by a 504 Plan are not as well described.

Post-secondary education and employment

Post secondary education is education after high school, such as university, community college, technical or vocational training. Here are some available resources:

DO-IT: DO-IT stands for Disabilities, Opportunities, Internetworking and Technology. A great resource for students with disabilities called DO-IT (Disabilities, Opportunities, Internetworking, and Technology) can be found at www.washington.edu/doi/. The DO-IT website provides information on the following topics and more: preparing for college, funding strategies (including information about scholarships), college survival skills, obtaining accommodations in hire education, using technology to maximize success in college and careers, a list of DO-IT programs, etc.

“Sarah Goes to College:” “Sarah Goes to College” is an informative and inspirational article about a young woman with FOP and how she finds the accommodations and financial support that she needs to attend college and live on campus. The story shows how determination, perseverance, and a long to-do list can bring you to your goal. It was

published in the IFOPA's 2004 newsletters. To read it, go to the following URL:
<http://depts.washington.edu/healthtr/notebook/transitionstories.pdf>

Disabled Student Services Office or DSS: Every college and university that accepts federal dollars (nearly all schools) has an office that works to ensure that students with documented disabilities receive the accommodations they need. Accommodations might include the following: note-taker, lab assistant, group lab assignments, classrooms moved to an accessible location, extra exam time, scanning services, computer with special assistive technology (such as on-screen keyboard, voice input, trackball mouse, etc.), and textbooks on CD.

The following is a list of resources available in the United States. Other countries may have similar resources.

Department of Vocational Rehabilitation, or DVR or VR: Vocational Rehabilitation is a program of the US federal government. It is administered by each state. VR's mission is to enable individuals with disabilities to obtain and keep employment. If a person qualifies, which is based on degree of disability and financial need, VR can pay for tuition and books, assistive technology (such as the most current and comprehensive computer hardware and software), mobility devices, transportation, hearing aids, attendant hours, and more to help a person get training for the kind of work he or she wants to do. Look for contact information in the telephone book under "State" or search for "Vocational Rehabilitation + (your state)" on the Internet.

Legal Rights: According to Section 504 of the Rehabilitation Act of 1973 and the Americans with Disabilities Act of 1990, "no otherwise qualified individual with a disability shall, solely by reason of his/her disability, be excluded from the participation in, be denied the benefits of, or be subjected to discrimination under any program or activity of a public entity." These are the rights of everyone with a disability everywhere in society.

Ticket to Hire: Ticket to Hire is a free national referral service to assist employers in hiring motivated, qualified workers with disabilities from the Social Security Administration's Ticket to Work Program. Ticket to Hire is a recruiting resource for employers that links employers to Employment Networks in their community that have job-ready candidates. You must be receiving services from an Employment Network or State Vocational Rehabilitation Agency in order to be referred for a job from Ticket to Hire. Employers, Employment Networks, and State Vocational Rehabilitation Agencies can learn more about TICKET TO HIRE by calling 866-TTW-HIRE, or 866-889-4473 (V/TTY) or E-mail at tickettohire@earnworks.com. Also, see www.earnworks.com.

Supplemental Security Income (SSI): SSI is a needs-based program that pays a monthly income to people with disabilities. Family income and assets are considered in determining whether a child qualifies for SSI. At age 18, a person may have up to \$2000 in assets and still qualify for SSI. Persons who receive SSI benefits also qualify for Medicaid Health Insurance and attendant care. The Social Security has other programs you may qualify for, including Social Security Survivors Benefits and Social Security Disability Benefits. For more information, please see www.ssa.gov or call 800-772-1213.

How to find helpful items

For more information on the subject of FOP and School, and ideas on possible adaptations, please see the following:

Chapter 25, “Finding Resources,” contains a list of items which may be helpful for school and life.

The IFOPA’s *Catalog of FOP Resources*, available at the IFOPA website at www.ifopa.org. See sections on “Electronics” for computer-related items, “Informational Resources” for information on children and youth, and “Education and Recreation.”

United States laws governing education of students with disabilities

In the United States, there are several laws that support the provision of special services for children with disabilities and they apply throughout the school experience, from kindergarten to graduation. The most cited law is the Individuals with Disabilities Education Act (IDEA), the federal law that requires public schools to make available to all eligible children with disabilities a free appropriate public education in the least restrictive environment appropriate to his or her individual needs. If children are found to be in need of special education services under this law, they receive an Individualized Education Plan, or IEP.

The second law which supports the inclusion of children with special needs is Section 504 of the Rehabilitation Act of 1973, the federal law that prohibits discrimination on the basis of disability in institutions receiving Federal funds, such as schools. This law requires a “level playing field” for all students, including those with disabilities. Children receiving services under this law receive a written “504 Plan”. For more information on these laws and various other issues of educating a child with special needs, please see www.wrightslaw.com, an invaluable website that provides easily understandable information for parents about

education, special education, advocacy and the successful management of the education bureaucracy.

Which of these laws best applies to the situation of any given child with FOP depends on several factors, including physical symptoms, academic performance, and the standards of the state in which the child lives. The school district may classify a student with FOP as in need of Special Education assistance under the IDEA because FOP is causing an “educational impairment” that only a complex assortment of services can accommodate, such as full-time aide, Physical & Occupational Therapy, Assistive Technology services, and/or door-to-door transportation. On the other hand, some school districts refuse to classify a child with FOP this way unless the child is performing below grade level, i.e. exhibiting an “academic” impairment.

In order to secure the necessary accommodations in the most efficient way, it is very important for the parents to get to know the people involved in the special education and disability field in their school district. The parent can seek advice from the teachers, principal, Special Education PTA representative in the school (if there is one), special education liaison at the school (such as the resource room teacher), and parents of other children with disabilities. Of course, it helps to know the state and Federal laws backing up your request for services and accommodations. The Wrightslaw website mentioned above (www.wrightslaw.com) has a state by state “yellow pages” that will give you local sources that may be able to advise you. *Exceptional Parent Magazine*, another great resource for parents of school age children with special needs and/or chronic illnesses, also has an excellent state by state resource guide. Please see www.EParent.com for more information. Whatever the parent’s level of special education knowledge, tact, mutual respect, and a desire to educate about FOP will go long way towards getting the child whatever he or she needs to succeed at school.

25.

Finding resources

By Sharon Kantanie

Resources for people with disabilities are thankfully much easier to find than in the days before catalogs and the world wide web. Yet, with the specialized needs of FOP, finding just the right gadget or tool isn't always easy. Here are some suggestions to make the search a bit easier.

One of the first resources many families today can turn to is other families living with FOP. Even though families are geographically separated from one another, help is just a phone call or an e-mail away. One way to reach many families all at once is through FOPonline, an e-mail newsgroup. It's much easier to see if someone has encountered the same challenge before than to reinvent the wheel and start from scratch. You can learn about unique inventions like an ice cream holder, an automatic page tuner, and a golf club fork, or learn how other families deal with such varied issues as schooling and whether it is okay to pierce one's ears. For more information about FOPonline, contact the IFOPA at 407-365-4194 or e-mail together@ifopa.org.

Another good resource is the IFOPA's Catalog of FOP Resources, available at the IFOPA website at www.ifopa.org. It is a database of tools and gadgets, including items used by members of the IFOPA as well as items carefully assembled from equipment catalogs, websites and word of mouth. If you do not have internet access, contact the IFOPA and they will be happy to help you.

What do you do if you still can't find what you are looking for? Well, then it helps to be resourceful. If you have access to the internet, go to your favorite search engine (such as Google). Think about words to describe what you are looking for. Often typing in a good search description is an important component in helping you find information quickly. If you are too vague, then you may get too many results, and the links that would be most helpful may not appear in the first pages. If you don't see what you need, try changing your words. Sometimes you might come across a link that isn't exactly what you need at the moment but seems interesting or might be something that could be helpful later. A truly resourceful person saves that information as well!

Another good approach is to ask questions of people whose jobs relate to your needs. Occupational therapists can be useful to people with FOP because they have access to information about special items that can help a person be more independent. A doctor can help you with a health issue, or maybe help you find a caregiver if you are looking for help. A

teacher's aide, in addition to providing assistance, might know of a tool that can provide greater independence.

Disability-related magazines are also a good resource. Consider keeping back issues—given the progressive nature of FOP, you never know when something you read months ago might prove useful. Also consider asking to be on the mailing list of a company that makes or sells products designed to help people with disabilities. At the end of this article is a list of resources.

Also, don't discount your own grey matter. Think about whether you might be able to solve the problem yourself, for example designing a gadget that doesn't yet exist. The outcome doesn't need to be sophisticated as long as it gets the job done. If you have an idea, but don't have the capabilities to build something yourself, seek out an engineer or a craftsman. One person with FOP did this and developed a bed that can be raised to a standing position. A mother had a special bicycle crafted for her daughter, adapting concepts she saw in other bicycles for children with special needs.

FOP often requires thinking "out of the box" and being creative. Each person will have different needs and different products and services are available in different countries. Hopefully this article has given you some starting points that will help you find the things you need. Keep reading to get more ideas about where to turn for help. The resources listed include information from IFOPA members across the globe, as well as information from the IFOPA's *Catalog of FOP Resources*, available at the IFOPA website at www.ifopa.org. (Go online for pictures and more information.).

Bedroom and bath

Item	Description	Where to find it
Mangar lifts, "Mangar Booster" and "Pillow Lift"	Transfer lifts	Tel: 01544 267674 in United Kingdom
Bed pull up strap	Straps that can be used to help a person move in bed	Available at medical supply stores
Cushions and mattresses	Many options are available.	See "Staying comfortable" in the "Miscellaneous topics from head to toe" in chapter 12. Also see IFOPA's <i>Catalog of FOP Resources</i> , available at www.ifopa.org .
Standing bed - option 1	Bed that can be raised to standing position	Logic move Stand-Up bed from www.auforum.com or international@auforum.com ; available in Europe
Standing bed - option 2	Bed specially adapted to be able to raise someone to a vertical position	www.ottobockus.com or 800-328-4058 (international company, bed adapted by a company in Germany)
Standing bed- option 3	Bed specially adapted to be able to raise someone to a vertical position	Contact the IFOPA for more information, including a newsletter article on this bed
Intercoms	Helpful for night time (or any time) communication.	Can use store bought intercoms or professionally install an intercom system. An alternative is to use a Panasonic PBX phone system like those used in offices. This is a very versatile option that can provide room-to-room or whole house paging, but it also more expensive and may require professional installation.
Faucet turner	Allows a person with limited reach to control the faucet	Home made with a hinge, dowel, and Styrofoam (See IFOPA's <i>Catalog of FOP Resources</i> , available at www.ifopa.org .)

Sink modification	Allows a person with limited reach to control the faucet	Installation of gooseneck faucet with separate controls that can be mounted at front of cabinet. (See IFOPA's <i>Catalog of FOP Resources</i> , available at www.ifopa.org .)
Kohler "Touchless Faucets" and Delta "e-Flow" faucet	Faucets recognizes when a person is near and turns on/off automatically	www.kohler.com or www.deltafaucet.com Contact a Kohler or Delta dealer in your area
Vitra wall mounted toilet	Can be mounted at the best height for an individual	www.vitra.com Contact a Vitra dealer in your area
Raised toilet seats	Clips onto toilets to raise height of seat	Contact a medical supply company in your area
Washlet	Toilet seat with integrated bidet (water wash)	www.washlet.com or contact Toto dealers
Clessence "Advanced Toilet Seat"	Bidet seat fits most toilets, features both posterior wash and feminine wash	www.ncmedical.com or 800-821-9319
Toilevator	Installs underneath a standard toilet, raises height 3 1/2 inches	www.ncmedical.com or 800-821-9319
Bathroom grab bars	For safety in the bathroom	www.ottobockus.com or 800-328-4058 (international company)
Roll-in showers	No threshold for easier entry	www.barrierfree.org , 877-717-7027, www.clarkmedical.com or 800-889-5295, Silcraft at 800-348-4848. Can also have a roll-in shower custom built (one inch unglazed tiles are suggested to prevent slipping)
Oximed	Bathroom adapters, special beds, mattresses, etc.	www.oximed.com.br

Computer items

Item	Description	Where to find it
Targus Wireless Multimedia Presenter	Wireless trackball with additional presentation features	www.targus.com
Trackballs	Wired and wireless options available, optional way to operate computers instead of using a mouse	Kensington (www.kensington.com) and Logitech (www.logitech.com) both make trackballs
Rollermouse Pro	Designed to be an ergonomic alternative to a mouse	www.rollermousepro.com Demo and online video available
On-screen keyboards	On-screen keyboards, typing using mouse movements, word prediction	www.lakefolks.org/cnt (for PC), www.ahf-net.com/reach.htm (for PC) or www.assistiveware.com/keystrokes.php (for Mac),
BAT keyboard	Keyboard can be operated with one hand	www.infogrip.com or 800-397-0921 (See IFOPA's <i>Catalog of FOP Resources</i> , available at www.ifopa.org .)
Ergonomic keyboards	Many companies make ergonomic keyboards.	www.ergonomicsmadeeasy.com
Speech recognition software	Use a computer by talking into a microphone	Naturally Speaking (PC) and Via Voice (PC and Mac) at www.nuance.com , iListen (Mac) at www.macspeech.com
Home controls	Voice activated computer systems can control almost anything in your environment	www.multimediasigns.com or 888-353-3996
Ergomart computer mount	Can raise, lower, and tilt the computer screen. Can be wall or desk-mounted	www.ergomart.com
Adjustable keyboard tray	Allows user to keep keyboard at adjustable heights, also tilts	Several companies make these: www.humanscale.com , www.neutralposture.com , www.ideaatwork.com

Abledata database	Database of equipment	www.abledata.com or 800-227-0216
RESNA (Rehabilitation Engineering & Assistive Technology Society of North America)	Technical assistance project	www.resna.org or 703-524-6686
National Council on Independent Living	Will evaluate your needs and help you locate funding	www.ncil.org or 877-525-3400

Cooking and eating

Item	Description	Where to find it
Personal blenders	Useful for those with fused jaws	See local stores that stock kitchen supplies.
Mini food processors/choppers	Useful for those with fused jaws	See local stores that stock kitchen supplies.
Sure Fit food guard	Attaches to plates, for those who need to scoop up their food to pick it up	www.ncmedical.com or 800-821-9319
Stir Chef	Automatically stirs pots	www.chefscatalog.com
Comfort Grip fork	Fork with extension, angled	www.ncmedical.com or 800-821-9319; item located only in "Functional Solutions" catalog and must be ordered by calling retailer directly
Golf club fork	Fork with extension	Home made with fork, golf club shaft cut down to size needed, and epoxy
Spork	Fork and spoon in one utensil	Home made - weld fork and spoon together
Rocker knives	Can be used with one hand	www.ncmedical.com or 800-821-9319
Swivel utensils	Utensils swivel and keep food level	www.sammonspreston.com or 800-323-5547
Flexible utensils	Feature flexible hands up to 14 inches long	www.sammonspreston.com or 800-323-5547
Long straws	Longer than average straws	Whistle Sippers from Abilitations 800-850-8602 or clear flexible plastic straws from www.ncmedical.com or 800-821-9319, www.sammonspreston.com or 800-323-5547

Spill-proof cups	Can be used while lying down	www.sammonspreston.com or 800-323-5547
Drink-Aide	Insulated water bottle with no spill feature	www.drink-aide.com or 800-336-7022
Items for eating	Adaptations for forks, spoons, etc.	www.expansao.com

Dressing and grooming

Item	Description	Where to find it
Long handled brush	Home made so that brush can be customized to meet an individual's needs	Home made with conduit pipe and a brush. Grind down brush to fit in pipe and secure with a screw. See IFOPA's <i>Catalog of FOP Resources</i> , available at www.ifopa.org for picture.
Long handled brushes and combs - option 2	handles up to 14 1/2 inch, options include a special brush for washing hair	www.ncmedical.com or 800-821-9319
Dressing stick	Long stick with different hooks on each end. Can be used creatively for many purposes.	Most medical supply stores should have this item. Also available through catalogs and the web. Folding/portable dressing stick available at www.ncmedical.com or 800-821-9319
Shoe horn with long, flexible handle	Can be used from a standing or seated position	Make Life Easier, www.make-life-easier.com at 800-522-0227
Sock aids	Useful for those with limited reach, Firmer models tend to work better than flexible ones	www.ncmedical.com or 800-821-9319
Elastic shoelaces	Elastic shoelaces stretch so that untying/tying may be unnecessary	www.ncmedical.com or 800-821-9319
Hair dryer stand	Height adjustable with flexible gooseneck	www.homefocuscatalog.com or 800-634-9585
Toothbrush holder	Home made extension	Home made, use a dowel rod, waterproof tape and if necessary an object to create space between the dowel and the toothbrush so that brush can fit in charger easily

Apparel for people with disabilities	Several companies design clothes for people with disabilities who may have special clothing needs	www.ableapparel.com www.adaptationsbyadrian.com www.rolli-moden.com www.makoa.org/clothing.htm www.supportplus.com www.special-clothes.com www.speciallyforyou.net www.wheelchairjeans.com
Coat modifications	Modifications that can be made by a tailor	Put a zipper in the back of a coat (doesn't work with hoods), have a tailor cut out a portion of the bulk in sleeves, replace under arm seams with Velcro—extend Velcro part of the way down the arm, slippery lining, particularly in arms, can make a coat easier to get into
Coat options	Store bought coats and capes	Look for coats with wide shoulders and deep armpits (Lands End can provide details on their coats), Thinsulate coats are warm without being bulky, Snowboard jackets have zippers/vents under the arms (Campmor catalog at 800-CAMPMOR has coats with these features), Capes can sometimes be found in local stores—also available from cascobaywoolworks.com or www.ebay.com
Items for shaving, brushing hair, etc.	Company also sells items for eating and wheelchairs	www.expansao.com

Home modifications

Item	Description	Where to find it
Open Sesame door opener	Opens and closes doors	www.opensesamedoor.com or 800-673-6911
Offset door hinge	Special hinges that add 2 inches to the width of a doorway, creating easier access for wheelchairs	www.dynamic-living.com 888-940-0605
Magic motion doorknob	Lets you open a door without turning a knob (Lever handles can also be helpful!)	www.dynamic-living.com or 888-940-0605
Motion activated light control by Leviton (occupancy sensor)	Motion activated light switch	Home Depot or www.leviton.com
Wall switch extender	Long handle is added to light switch to make it easier to control for those with limited reach	www.dynamic-living.com or 888-940-0605
Kid's switch	Glow in the dark switch extender	www.leapsandbounds.com 800-477-2189
"Anywhere Switch"	Remote light and appliance control	Home Depot or www.leviton.com
Home automation	Home automation can involve automating lights, whole house video distribution (so you don't need to manually insert DVDs, CDs, etc.), and almost anything else you can imagine	Contact a company that specializes in home automation.
QRO Systems "Home Manager"	Control doors, TV, video, phone, appliances, etc. by remote control	01 473 212218 in United Kingdom
Wheelovator home lift	Lift can allow for easy entry into home	www.wheelovator.com or 800-968-5438

Home elevators	Available in various sizes. If you need to install one, you may want to consider making it oversized for easier access for power wheelchairs.	Contact a company that installs lifts and elevators in your local area.
Pull down closet rod	Closet rod drops down to be more accessible	www.organizeit.com or 800-210-7712
AD-AS height adjustable sink, cabinets, cook top	Can be raised or lowered with a touch of a button	www.ad-as.com or 800-957-2720
Idea Center	Collection of products from companies that emphasize universal design and accessibility	www.ap.buffalo.edu/idea/Brightideas/
Wheelchair.net Community Living Links	Collection of products that emphasize universal design and accessibility	www.wheelchairnet.org/WCN_Living/homemod.html
INFINITEC (Infinite Potential Through Assistive Technology)	Collection of products that emphasize universal design and accessibility	www.infinitec.org/live/homemodifications/basics.htm
Disability Resources	Collection of websites that are helpful in building or adapting a home	www.disabilityresources.org/ARCHITECTURE.html
National Directory of Home Modification Resources	Lists home modification resources in the United States	www.usc.edu/dept/gero/nrcshhm/directory

Office and school

Item	Description	Where to find it
Slant boards	Angle adjustments allow a person to work at any angle.	see www.tifaq.org/accessories/slantboards.html for numerous options. Healthbydesign.com makes a model that can go from sit-to-stand. Slant boards or reading boards can be found in many catalogs of products for people with disabilities.
Chalkworks chalk writer	Made by Cadaco, 11 inches long, also available in 18 inch length, holds chalk	Bed, Bath and Beyond
Intellitools	Mathpad provides a way for children who have difficulty writing to solve math problems on-screen	www.intellitools.com or 800-547-6747
Ergoquest workstations	Computer desks that use a table that can work with a recliner or be used in bed	www.ergoquest.com or 888-298-2898
Height adjustable work stations	Adjust the height using either a crank or electronically, some have a second layer at a different height for monitor or keyboard	www.ergonomicconnection.com
Stance Angle chair	Chair can be adjusted into many positions, including a reclined standing position	www.healthpostures.com or 800-277-1841
Stand up chair	Allows a person to work standing with some support	www.posturite.co.uk
Wheelchair backpacks	Numerous options. Over-the-chair strap (slip strap) can be used for scooters and power wheelchairs	www.advantagebag.com or 800-556-6307
Adjustable work surface	Work surface can be adjusted to needed slant/height	www.easethepainllc.com

Family Village	Online resource, school section contains many links to educational resources	www.familyvillage.wisc.edu
Kid Source	Information for parents of children with disabilities regarding education issues	www.kidsource.com
Whizz Kidz	Organization to help children with disabilities at home and school, based in United Kingdom	www.whizz-kidz.org.uk

Reaching

Item	Description	Where to find it
Scratching hand/Telescoping back scratcher	Scratching hand is useful for extending reach to adjust glasses, scratch an itch, reach objects, etc.	www.shophometrends.com or 888-815-0814
Reachers	Numerous options are available, both in terms of length and style	Should be available at medical supply stores or www.ncmedical.com or 800-821-9319
Dressing stick	Useful for extending reach and for other purposes too.	Should be available at medical supply stores or www.ncmedical.com or 800-821-9319

Recreation

Item	Description	Where to find it
Playing card holders and automatic shufflers	Make playing cards easier	www.thehouseofcards.com/card_accessories.html
Ice cream holders	one holds ice cream on sticks, one holds cones	
Compagnie page turner	Electronically turns book pages	www.handisurf.net/tourne-pages-compagnie/index.htm
Book holders	Holds books to lessen strain of holding a book or looking at an awkward angle	Many catalogs of products for people with disabilities feature book holders, May also find locally
Specialty bicycles	Adaptive bikes for use by people with a variety of special needs, most will need handlebar modification for use by someone with FOP	www.rocknrollcycles.com www.freedomconcepts.com www.haverich.com , www.trailmate.com
Folding bicycle	Bicycle was modified to individual's needs by a craftsman	If specialty bikes are unavailable in your country, show a craftsman bikes you like and ask him to design something
Bicycle adaptations	Altering handlebars and adding training wheels	Contact a local biking store for ideas on altering handlebars and adding training wheels to improve balance
Embroidery stand	Do needlepoint without needing to hold your work	Contact local needlepoint store, www.levencrafts.co.uk www.stitchers-paradise.com

Other resources - International

Item	Description	Where to find it
Association Française contre les Myopathies (AFM)	French association for neuromuscular diseases, supports patients (France)	www.afm-france.org or +33.1.69.47.28.28
Identités	catalog of products (France)	www.identites.tm.fr
Associaton des Paralysés de France	French association for people with disabilities (France)	www.apf.asso.fr
Handisurf.net	Website for people with disabilities (France)	www.handisurf.net
Fondation Garches	Website for people with disabilities, features many resources and a list of products and services (France)	www.handicap.org
T.A.C.Maides Techniques	Catalog of products (France)	+33.2.47.05.73.29
French resources	Learn about disability pensions, care assistants, and funding for equipment	CCAS at local town halls or Maison du Handicap
Handicat	Information (France)	www.handicat.com
Search tool for items	Search for available items to help people with disabilities	www.aides-techniques-cnsa.fr
CIZ	Resource for the Netherlands	www.ciz.nl
Ableize	List of disability-related resources (United Kingdom)	www.ableize.com
Disability UK	List of disability-related resources (United Kingdom)	www.disabilityuk.com

Persons with Disabilities	Guide for people with disabilities (Canada)	www.pwd-online.ca
Enablelink	Guide for people with disabilities (Canada), also published Abilities magazine.	www.enablelink.org

Other resources - United States

Item	Description	Where to find it
Disability Resources	Website with information on numerous disability-related topics	www.disabilityresources.org
Bandaides and blackboards	Site with information for children, teens, and adults.	www.lehman.cuny.edu/faculty/jfleitas/bandaides
Special Needs Advocate for Parents (SNAP)	Advocacy, estate planning, etc. Publishes a newsletter with information on various topics	www.snapinfo.org
Sammons Preston	Catalog/website with disability-related items	www.sammonspreston.com or 800-323-5547
Dynamic Living	Catalog/website with disability-related items	www.dynamic-living.com or 888-940-0605
Functional Solutions	Catalog/website with disability-related items	www.ncmedical.com or 800-821-9319
United States Resources	Centers for Independent Living, Vocational Rehabilitation, Medicaid services, Supplemental Security Income	Resources may vary by state
Medication resources	Resources for helping to pay costs of medications	www.freemedicine.com www.rxoutreach.com
Exceptional Parent	Monthly magazine for parents of children with special needs	www.eparent.com or 800-372-7368
New Mobility	Magazine about disability-related issues	www.newmobility.com
National Family Caregivers Association	Non-profit association for caregivers	www.thefamilycaregiver.org
Council for Disability Rights	Organization started by a mother of a child with FOP, website features resources and information on schooling, home modifications, and more	www.disabilityrights.org

See Chapter 4, “FOP and mobility,” Chapter 13, “Miscellaneous health topics from head to toe,” and Chapter 24, “FOP and school: Ideas and resources” for more information.

26.

IFOPA services

The IFOPA helps families meet the challenges of living with FOP.

Introduction

The International FOP Association, or IFOPA, is a nonprofit 501(c)(3) charitable organization supporting families coping with the rare genetic condition called fibrodysplasia ossificans progressiva (FOP). The IFOPA was established by Jeannie Peeper, a woman with FOP, in 1988 as a means to end the isolation brought upon by FOP. The IFOPA's mission is to instill hope through research, education, and support while searching for a cure for FOP.

The IFOPA's goals cannot be accomplished without the help of several remarkable groups of people: our wonderful volunteers, who graciously give their time and talents; the FOP families, who actively fundraise; our gracious and generous contributors; our extraordinarily dedicated research team; our generous Board of Directors, which leads the organization in both its long-term and short-term goals; and the small IFOPA staff, who works so hard and cares so much. Together, we are an incredible team. We hope that you will join us in our mission to find a treatment and cure for FOP.

To date, the IFOPA has donated more than \$5 million in support of a treatment and cure to the FOP research effort at the University of Pennsylvania School of Medicine. Currently, the IFOPA contributes approximately \$500,000 annually to meet the FOP Lab's needs and to help realize the hope for a treatment and cure.

The IFOPA dedicates itself to helping families meet the challenges of living with FOP by providing the following services.

Website

The IFOPA maintains a website at www.ifopa.org, which provides both IFOPA members and the general public with accurate information about FOP, families affected by FOP, and developments in medical research.

IFOPA newsletters

The *FOP Connection* is the IFOPA's quarterly newsletter. It features articles and pictures about people with FOP, fundraising events, research developments, and suggestions and resources for living with FOP. *Milestones* is a semi-annual newsletter for donors that focuses on research developments.

Betty Anne Laue/IFOPA Resource Center

The Betty Anne Laue/IFOPA Resource Center was named in memory of a dear friend of the FOP community, former Board Member and grandmother of FOP member Ian Cali. The Resource Center is a central clearinghouse for information on FOP. It offers scientific and medical articles, medical binder kits for collecting personal medical information, emergency cards with basic information about how to handle emergencies and whom to contact (see chapter 5 for more information about both the medical binder kits and emergency cards), interesting stories on FOP members and fundraisers, videos, and more. Most of the materials in it are supplied free of charge, though there is a nominal fee for the videos. For a list of all available resources, including print copies of *What is FOP? A Guidebook for Families* and *What is FOP? A Guidebook for Children*, contact the resource center using the information below:

Digital versions of both guidebooks are available on the IFOPA website at www.ifopa.org, in the "Living with FOP" section.

IFOPA
 P.O. Box 196217
 Winter Springs, FL 32719-6217
 Telephone: 407-365-4194
 Fax: 407-365-3213
 Website: www.ifopa.org
 E-mail: together@ifopa.org

FOPonline e-mail newsgroup

The IFOPA encourages its members to join FOPonline, an e-mail newsgroup. It gives you the opportunity to share your thoughts, concerns, and questions. It is important for the FOP community to keep the lines of communication open because we all learn from one another only if we share our discoveries and express ourselves. The electronic mailing list is a way to bridge the gap created by the miles which usually separate us from one another.

Below are some commonly asked questions about mailing lists. If you have additional questions, or if you would like to join the list, please contact the IFOPA at 407-365-4194 or together@ifopa.org.

What is an e-mail mailing list?

A mailing list is an automated method of delivering messages to all people subscribing to an e-mail list on a given topic. When you post a message, it is immediately sent to everyone on the list. Mailing lists are one of the most effective ways to use the internet for group discussions.

What kind of information can a person expect to find on FOPonline?

FOPonline's main goal is to create an online community where people can post questions and get advice and support from other families and individuals interested in FOP.

Who can join FOPonline?

Enrollment in FOPonline is open to individuals with FOP, family members, medical professionals, or a person with some other personal connection to someone with FOP. When joining, please note your name, phone number, and relationship with the FOP community.

What about privacy?

Privacy is an important issue. The FOP mailing list is set up so that the list of members is not available to participants. If you do not post messages, then only list administrators will have access to your name and e-mail address. However, when you post a message, participants will see your e-mail address.

If you speak Spanish or Portuguese, you may wish to join FOPlatinoamerica (Spanish) or FOPbrasil (Portuguese). For information, contact info@fundacionfop.org.ar to learn more about FOPlatinoamerica or sosfop@uol.com.br to learn more about FOPbrasil.

The FOPonline newsgroup guidelines are posted on the IFOPA website at www.ifopa.org in the "Members Center" section.

We also want to let you know about Pray for FOP Healing, an online group that gives IFOPA members an opportunity to share prayer requests, inspirational quotes, and poems, and to talk about how faith sustains them. For more information or to join, contact Theresa Caruso at hamsancity@aol.com. While this group is part of IFOPA services, The International FOP Association (IFOPA) is a non-denominational/non-religious organization, and, as such, it does not endorse, serve, or favor any specific religious organization, practice, sect, or idea of any kind.

The Quality of LIFE Award

The IFOPA's program that provides information about disability rights laws, education, adaptive equipment, assistive technology, government entitlements, advocacy and more is called LIFE. LIFE stands for "living independently with full equality."

In addition to providing advice and information, through special funding the IFOPA offers the Quality of LIFE Award. The Quality of LIFE Award was developed to assist IFOPA members in living more independently in their daily lives.

Here is more information about how the Quality of LIFE Award works:

- LIFE Awards are for IFOPA members who have paid their annual dues or who have asked to be sponsored and have provided verifiable medical confirmation of the FOP diagnosis to the IFOPA.
- There are no guarantees that a request will be granted. The decision is made by a committee, and awards will be given in the order applications are received until funds for the program are used up.
- The IFOPA maintains a LIFE Award Fund to pay for this grant program. The money for this fund comes from donors and family fundraisers who direct their donations to the LIFE program. Donors and family fundraisers are encouraged to contribute to the LIFE Award Fund so that LIFE Awards can continue to be provided to IFOPA members.

To be considered for a LIFE Award, please contact the IFOPA for an application or download an application from the IFOPA website at www.ifopa.org in the "Member Center" section.

For more information about the LIFE program or the Quality of LIFE Award, contact the IFOPA at 407-365-4194 or together@ifopa.org.

Family meetings

The IFOPA believes in the importance of bringing families dealing with FOP together as a means to share information and support one another. For this reason, the IFOPA tries to provide the rare opportunity for families from all over the world to meet in one place, as well as get appointments with Dr. Fred Kaplan, Dr. Eileen Shore, dentists, orthodontists, and other medical professionals so they can discuss their medical concerns about FOP. We have been very fortunate to obtain funding for our family meetings, which have been held in 1991, 1993, 1994, and 2004, respectively.

International FOP symposia

Building awareness and support across the globe, the IFOPA's international symposia enable families, scientists, and medical specialists to come together to share knowledge and to exchange ideas and experiences. Unlike the organization's family meetings, symposia are greater in scale and scope and often feature a larger number of attendees from around the world. The IFOPA has hosted four international symposia on FOP, in 1991, 1995, 2000, and 2007, respectively.

Making it all happen

Every day, the FOP researchers move closer to finding a way to deter the growth of FOP bone. In order to speed up the investigation into possible treatments, the IFOPA has committed itself to insuring a constant flow of funds to the FOP Research Laboratory at the University of Pennsylvania School of Medicine.

In addition, the IFOPA continues to help those with FOP in other ways. It supports member conferences, educates the general and medical communities, connects members through newsletters and its website, assists with fundraisers, and addresses patient needs. Without the dedication and commitment of compassionate supporters, the IFOPA could not fulfill its mission.

Family fundraising support

Please consider getting involved through fundraising. It can be tons of fun and the best way to insure a bright future for the one in your life with FOP. Because of the organization's grass roots mindset, fundraising ideas of all kinds are acceptable, from large scale events to intimate dinners at home. Remember: Every fundraiser is important, big, small and in-between.

Additionally, the IFOPA can assist you with your fundraising efforts, providing necessary informational materials such as *Milestones* newsletters, IFOPA/FOP "Facts in Brief" sheets, videos, brochures, donation envelopes, awareness merchandise and planning assistance. For more information, including written outlines for certain events and a list of possible fundraiser ideas, or if you have any fundraising questions, please contact the IFOPA at 407-365-4194 or together@ifopa.org.

IFOPA accomplishments

If you would like to review the many accomplishments the IFOPA has achieved over the years, please visit the organization's website at www.ifopa.org.

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FOP resources around the world

For more information about FOP, you may contact the following organizations, visit numerous websites, or join an e-mail newsgroup.

International FOP Association or IFOPA

P.O. Box 196217

Winter Springs, FL 32719-6217

Telephone: 407-365-4194

Fax: 407-365-3213

Website: www.ifopa.org

E-mail: together@ifopa.org

Fundación FOP (Argentina)

Bonpland 1964 - Dto 3 PA - (1414) Ciudad Autónoma de Buenos Aires
Argentina

Telephone: 54-11-9-15-4145-5391 or 54-11-4771-7927

Fax: 54-11-4771-7927

Website: www.fundacionfop.org.ar

E-mail: info@fundacionfop.org.ar

FOP Brasil (Brasil/Brazil)

Rua Pedro de Toledo 129 cj 121 Vila Clementino

Cep – 04039-030

São Paulo – SP

Brazil

Telephone: 55-11-5539-5817

Fax: 55-11-5539-5817

Website: www.fopbrasil.com.br

E-mail: sosfop@uol.com.br

Canadian FOP Families and Friends Network (Canada)

101 Brixham Circle

London, ON N6K 1K9, Canada

Telephone: 519-641-5742

Website: www.cfopn.org

E-mail: CanadianFOPNetwork@live.com

FOP-e.V. (Germany)

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Websites about FOP

The following websites were set up by individuals and families to help spread information about FOP:

Colombia

Website: www.geocities.com/alafof/principal.htm

E-mail: iag1979@hotmail.com

India

Website: www.fopinfo.blogspot.com

E-mail: 2005.lakshmi@gmail.com

Japan

Website of FOP Step Jump: www17.ocn.ne.jp/~j-fop

E-mail: kyozenm3@mbn.nifty.com

Poland

Website: www.fop.prv.pl

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E-mail-based support

In addition to the IFOPA's FOPonline e-mail newsgroup group (see Chapter 26, "IFOPA services," for more details), families may wish to turn to these resources.

FOPbrasil

FOPbrasil, based in Brazil, is an e-mail discussion group for FOP members who speak Portuguese. For more information or to join, contact sosfop@uol.com.br.

FOPlatinoamerica

FOPlatinoamerica, based in Argentina, is an e-mail discussion group for FOP members who speak Spanish. For more information or to join, contact info@fundacionfop.org.ar.

Pray for FOP Healing

Pray for FOP Healing is an online group that gives IFOPA members an opportunity to share prayer requests, inspirational quotes and poems and talk about how faith sustains them. For more information or to join, contact Theresa Caruso at hamsancity@aol.com. *While this group is part of IFOPA services, The International FOP Association (IFOPA) is a non-denominational/non-religious organization, and, as such, it does not endorse, serve, or favor any specific religious organization, practice, sect, or idea of any kind.*

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