



2012

Caring for Individuals with Fibrodysplasia Ossificans Progressiva (FOP): A Treatment Manual for Occupational Therapists

Amanda Rositas
University of North Dakota

Kathryn Zavoral
University of North Dakota

[How does access to this work benefit you? Let us know!](#)

Follow this and additional works at: <https://commons.und.edu/ot-grad>



Part of the [Occupational Therapy Commons](#)

Recommended Citation

Rositas, Amanda and Zavoral, Kathryn, "Caring for Individuals with Fibrodysplasia Ossificans Progressiva (FOP): A Treatment Manual for Occupational Therapists" (2012). *Occupational Therapy Capstones*. 159. <https://commons.und.edu/ot-grad/159>

This Scholarly Project is brought to you for free and open access by the Department of Occupational Therapy at UND Scholarly Commons. It has been accepted for inclusion in Occupational Therapy Capstones by an authorized administrator of UND Scholarly Commons. For more information, please contact und.common@library.und.edu.

CARING FOR INDIVIDUALS WITH FIBRODYSPLASIA OSSIFICANS
PROGRESSIVA (FOP): A TREATMENT MANUAL FOR OCCUPATIONAL
THERAPISTS

By

Amanda Rositas, OTS and Kathryn Zavoral, OTS

Advisor: Dr. Mandy Meyer, Ph. D.

A Scholarly Project

Submitted to the Occupational Therapy Department

of the

University of North Dakota

In partial fulfillment of the requirements

for the degree of

Master's of Occupational Therapy

Grand Forks, North Dakota

May 12th, 2012

This Scholarly Project Paper, submitted by Amanda Rositas, OTS and Kathryn Zavoral, OTS in partial fulfillment of the requirement for the Degree of Master's of Occupational Therapy from the University of North Dakota, has been read by the Faculty Advisor under whom the work has been done and is hereby approved.

Faculty Advisor

Date

PERMISSION

Title Caring for Individuals with Fibrodysplasia Ossificans Progressiva
(FOP): A Treatment Manual for Occupational Therapists

Department Occupational Therapy

Degree Master's of Occupational Therapy

In presenting this Scholarly Project/Independent Study in partial fulfillment of the requirements for a graduate degree from the University of North Dakota, I/we agree that the Department of Occupational Therapy shall make it freely available for inspection. I/we further agree that permission for extensive copying for scholarly purposes may be granted by the professor who supervised our work or, in his/her absence, by the Chairperson of the Department. It is understood that any copying or publication or other use of this Scholarly Project/Independent Study or part thereof for financial gain shall not be allowed without my/our written permission. It is also understood that due recognition shall be given to me/us and the University of North Dakota in any scholarly use which may be made of any material in our Scholarly Project/Independent Study Report.

Signature_____ Date_____

Signature_____ Date_____

TABLE OF CONTENTS

ACKNOWLEDGMENTS.....	iv
ABSTRACT.....	v
CHAPTER	
I. INTRODUCTION.....	1
Statement of the Problem.....	1
Purpose of the Study.....	3
II. REVIEW OF LITERATURE.....	10
History.....	11
III. METHOD.....	30
IV. PRODUCT.....	40
V. SUMMARY.....	105
APPENDIX A: Interview Consent Form.....	106
APPENDIX B: Product Content Usage Agreement.....	108
REFERENCES.....	110

ACKNOWLEDGEMENTS

The authors wish to thank Dr. Meyer for her patience, encouragement, and her time commitment for helping us through this process. Also, we would like to thank Kaylee Fenton for allowing us to use her personal story to guide the development of this project and the time she took to talk with us. We would like to thank our family members and friends for their support and love. We couldn't have completed this without the help from everyone and their support!

ABSTRACT

Fibrodysplasia Ossificans Progressiva is a rare and disabling genetic disorder that results in ossification of the soft tissues (Kaplan, Xu, et al., 2008; Kocyigit, Hizil, Hemis, Sabah, & Memis, 2001). There is a lack of research and literature on therapeutic intervention for those with a diagnosis of FOP. However, occupational therapists (OTs) can help those with FOP increase their quality of life through enhancement of activities of daily living (Kaplan, Le Merrer, et al., 2008). Overall, this product will serve as a guide for occupational therapists working with individuals diagnosed with FOP. The benefits of this product will be determined by the number of individuals who display interest in the use of this scholarly project.

The scholarly project is presented in the following four chapters. Chapter II presents the professional literature review regarding the definition and treatment recommendations relating to FOP. Chapter III will present the methodology of how the information from the literature review was obtained as well as education to develop the treatment manual. Chapter IV presents the actual treatment manual titled *Caring For Individuals With Fibrodysplasia Ossificans Progressiva (FOP): A Treatment Manual For Occupational Therapists*. Chapter V provides the conclusion, recommendations, and limitations of the scholarly project.

The limitations of the product include the lack of evidence-based literature specific to occupational therapy and FOP. Another limitation is that FOP is a rare genetic disorder and not many occupational therapists will work with individuals diagnosed with FOP.

The product may be implemented by adapting the product as a guide for occupational therapists treating individuals with FOP through a continuing education workshop. Occupational therapists that are currently working with individuals who are diagnosed with FOP can also utilize this product.

Overall, this product will serve as a guide for OTs working with individuals diagnosed with FOP. The benefits of this product will be determined by the number of individuals who display interest in the use of this scholarly project.

It is recommended that further research be done regarding occupational therapy treatment for individuals with FOP. More evidence-based research is needed to ensure quality treatment is provided for individuals diagnosed with FOP. Other suggestions to further the usefulness of this product include publication of information regarding OTs working with individuals diagnosed with FOP.

CHAPTER I

INTRODUCTION

Fibrodysplasia Ossificans Progressiva (FOP) is a genetic disorder in which muscle tissue and connective tissue are progressively replaced by bone; the disease produces bone outside the skeleton that eventually constrains movement (Parker & Parker, 2007). It is one of the most debilitating conditions of ossification of extraskeleton known to humans (Kaplan, Le Merrer et al., 2008). This disease is a rare genetic disorder that affects approximately 1 in 2 million people worldwide. There are only 700 confirmed cases of FOP across the globe and of those 700 cases, 185 known cases are in the United States of America. The disease has no predisposition to ethnic, racial, or religious affiliations (IFOPA, 2009b). Even though this is a rare disorder, OTs are in an excellent position to address all areas of occupation that are needed for a client diagnosed with FOP.

Kaplan, Le Merrer, et al., (2008) provides information regarding the medical treatment of FOP in the article *Fibrodysplasia Ossificans Progressiva*. The authors provided guidelines for the following areas: anesthesia, falls, flare-ups, hearing, immunizations, influenza, IVs, limb swelling, occupational therapy, physiotherapy, pulmonary function, school, surgery, and teeth (Kaplan et al, 2008). The International Fibrodysplasia Ossificans Progressiva Association (IFOPA) provides medical treatment guidelines, which include: avoidance of deep tissue trauma, avoidance of intramuscular

injections, protection of the neck and jaw during intubation, and consulting expert doctors regarding any risks that must be considered before medical interventions (IFOP, 2009a).

Although there are various resources on the treatment of individuals with FOP, the literature review provided limited information regarding the unique contribution from occupational therapy with this disease. The information the authors were able to gather from the literature review includes the definition and common symptoms of FOP, the importance of early diagnosis, the significance of trauma, psychosocial effects and the opportunity for genetic research.

Fibrodysplasia ossificans progressiva results in a progressive loss of range of motion, thus activities of daily living may be more difficult. Occupational therapists can help individuals afflicted with this condition live a more fulfilling life (Kaplan, et al 2008a). Occupational therapists are in an excellent position to address all areas of occupation that are needed for a client diagnosed with FOP.

The lack of literature regarding guidelines for an OT working with an individual diagnosed with FOP lead the authors to develop a treatment manual for that purpose. The manual consists of information for OTs to use as an educational resource when treating an individual diagnosed with FOP.

Model

The Model of Human Occupation (MOHO) is the theoretical basis for the derived product. Through the use of this model all aspects of an individual are considered including volition, habituation, and performance capacity. These aspects of an individual were considered with the development of the product. Also, the manual includes insight from an interview with an individual who is living with FOP to gain a perspective of the disease and explore how occupational therapy could assist.

Conclusion

The scholarly project is presented in the following four chapters. Chapter II presents the professional literature review regarding the definition and treatment recommendations relating to FOP. Chapter III presents the methodology behind the literature review and development of the treatment manual. Chapter IV presents the treatment manual titled *Caring For Individuals With Fibrodysplasia Ossificans Progressiva (FOP): A Treatment Manual For Occupational Therapists*. Chapter V provides the conclusion, recommendations, and limitations of the scholarly project. Overall, this project was completed to help OTs provide quality care to those with diagnosed with FOP. The next chapter consists of the literature that was utilized for the development of the treatment manual for OTs.

CHAPTER II

REVIEW OF LITERATURE

Introduction

A literature review was completed on Fibrodysplasia Ossificans Progressiva (FOP). The literature review consisted of the definition of FOP, and the prevalence, problems, and progression related to the disease. Additionally, the role of occupational therapy and what the profession has to offer for those diagnosed with FOP was reviewed. Occupational therapy's role in genetic research, early intervention, fall prevention, education, mobility, psychosocial issues, assistive technology, and home modification aspects related to occupational therapy are addressed in this literature review.

What is Fibrodysplasia Ossificans Progressiva

Fibrodysplasia Ossificans Progressiva is a genetic disorder in which muscle tissue and connective tissue are replaced by bone progressively; the disease produces bone outside the skeleton that increasingly constrains movement (Parker & Parker, 2007). It is one of the most debilitating conditions of ossification of extraskeleton known to humans (Kaplan, Xu, et al., 2008). Earliest documentation of this disorder dates back to the 17th and 18th centuries and was formally known as Myositis Ossificans Progressiva (International Fibrodysplasia Ossificans Progressiva Association [IFOPA], 2009f). In the 1970s, the name was changed to FOP to recognize that other soft tissues in addition to muscle were replaced by bone. It is now accepted that FOP affects the following types of

soft tissues: aponeuroses, fascia, ligaments, tendons, and the connective tissue of skeletal bone, turning them into mature bone (IFOPA, 2009f). FOP is a debilitating disorder that has no bias to who it affects.

Prevalence

FOP is a rare genetic disorder that affects approximately 1 in 2 million people worldwide. The disease has no predisposition to ethnic, racial, or religious affiliations (IFOPA, 2009b). There are only 700 confirmed cases of FOP across the globe and of those 700 cases, 185 known cases are in the United States of America. Of the 185 known cases in the United State of America, the skeleton of Harry Eastlack has been studied greatly by researchers in Philadelphia, Pennsylvania (IFOPA, 2009e).

The skeleton of Harry Eastlack (1933-1973), diagnosed with FOP, has been on display at The Mutter Museum of The College of Physicians in Philadelphia, Pennsylvania since his death (IFOPA, 2009e). Extra layers of bone caused by flare-ups of FOP connected his skeleton. Thus, Eastlack's skeleton is almost completely fused into one piece (Kaplan, 2005). Flare-ups begin as painful, warm, fibrous masses in soft connective tissue. Most often these masses progress through an endochondral pathway turning into mature bone (Kaplan, Shore, Glaser, Emerson, 2003). Through studying Harry Eastlack's skeleton, researchers have been able to identify and confirm many aspects of the disease including location of the gene affected by FOP (Kaplan, 2005).

Through genetic exploration, researchers have determined that a mutation on the ACVR1 gene causes FOP (Parker & Parker, 2007). This gene helps control growth and

development of bones and muscles. Scientists believe that the mutated ACVR1 gene is always active and causes the overgrowth of bone and fusion of joints (Parker & Parker, 2007). By locating the gene that is known to cause FOP there is now a strong clinical background for the diagnosis and prognosis of this disease as well as more well defined research into its cause (Smith, Anhanasou, & Vipond, 1996). Researchers are still conducting genetic studies to develop treatments that will prevent, stop, or even reverse the progression of FOP (Kaplan, Shen, et al., 2008).

Most individuals diagnosed with FOP result from new mutations in the gene and there is no history of the diagnosis in their family (Parker & Parker, 2007). The gene mutation is part of an autosomal dominant pattern where one copy of the altered gene in each cell is sufficient to cause FOP (Parker & Parker, 2007).

Definitive genetic testing is available to determine if an individual has FOP before the occurrence of heterotrophic ossification (Kaplan , Le Merrer, et al., 2008). If a clinician suspects an individual has FOP due to the malformation of the big toe or episodic soft tissue swellings, common symptoms of the disorder, he or she can perform these diagnostic procedures and avoid the harmful procedures that could cause heterotrophic ossification to occur (Kaplan et al, 2008a).

Progression

At birth, those with FOP often present with a malformed great toe, a symptom displayed by individuals with classical FOP. The malformed big toes are often short, bent, and curved inward (IFOPA, 2009d). Even though those with FOP have the disease

at birth and are often born with a malformed great toe, the supplemental bone does not start forming until childhood or later in life.

For example, in a case report by Kartal-Kaess et al. (2010) a patient was born with malformed great toes, but did not experience restricted mobility and an impaired gait until 13 years of age. She was seen by an orthopedic surgeon and a diagnosis of myositis ossificans was made without the consideration of the malformed great toe. Following diagnosis, she underwent a surgical procedure that removed the ossification of the left hip and experienced no pain and normal hip function for 3 months. However, recurrence of heterotopic ossification of the left hip was confirmed a year later and the patient's diagnosis was reconsidered. FOP may have been diagnosed before the invasive and harmful surgical procedure if the malformed great toe would have been taken into account with the ossification of the hip.

In another study of 16 patients (31 feet) diagnosed with FOP, deformity of the great toes was examined for the potential of identifying characteristics for early diagnosis (Nakashima et al. 2010). As a result of the study, 29 of the 31 feet displayed great toes that had several degrees of deformity. Also, 20 of the 31 feet presented a shortened great toe. However, only two feet of one patient presented with no obvious deformity of the great toe. Thus, a shortened deformed great toe is typically found on those diagnosed with FOP. Besides the malformed great toe present at birth, other symptoms of FOP present themselves at various times in an individual's life.

Typically, the disease starts to become noticeable in the first decade of life. The

symptoms often begin in the neck and shoulder and then proceed to the back, trunk, and the limbs (IFOPA, 2009d; Parker & Parker, 2007). The disease tends to advance in patterns. In a case study by Cohen et al. (1993) forty-four individuals with FOP were surveyed and patterns of heterotopic ossification were identified. FOP progressed from proximal to distal, upper extremities to lower extremities, and axial to appendicular. By the age fifteen, over 95% of the individuals surveyed had severe loss of mobility of the upper limbs.

The new bone formation progresses at different rates from person to person. These first symptoms consist of inflamed swellings on the neck, back, or scalp (IFOPA, 2009d). Scalp nodules (swellings) may be another significant factor in the diagnosis of FOP. In the study by Piram et al. (2011) a medical chart review showed that of the 43 medical cases evaluated all patients were diagnosed with FOP, all had deformed toes, and 17 had scalp nodules. Thus, scalp nodules, in addition to deformed great toes are some of the characteristics and early signs of FOP. These swellings may go away, but new bone is left behind. Besides the disease's natural progression, other factors can cause ossification as well.

Any trauma, such as immunizations or falls can cause harm to the muscles or soft tissue within the affected area, resulting in rapid ossification of tissue (Parker & Parker, 2007). Also, viral illnesses such as influenza may cause flare-ups of the disease and ultimately cause new bone formation (Kartal-Kaess et al, 2010; Parker & Parker, 2007). Other than trauma and viral illness, formation of bone most often occurs spontaneously

Cohen et al. (1993). This extra bone formation leads to a decrease in mobility and range of motion.

Overtime, those affected by the disease exhibit a progressive loss of mobility because FOP affects their joints (IFOPA, 2009c). Specifically, those with FOP may experience malnutrition due to difficulty eating and inability to fully open their mouth (Parker & Parker, 2007). In a case report by Herford and Boyne (2003), a 24-year-old male was unable to open his mouth for 10 years resulting in lack of dental care. The temporomandibular joint (TMJ) is typically the last joint to be affected and is symptomatic in 71% of individuals affected by FOP (Herford & Boyne, 2003; Wadenya, Fulcher, Grunwald, Nussbaum, & Grunwald, 2010). Those with TMJ symptoms may display poor oral hygiene, experience difficulty with dental care, and are at risk of vomit aspiration. Thus, preventive oral healthcare are crucial for those with FOP (Wadenya et al., 2010).

In addition to difficulties with the TMJ, breathing difficulties may be an issue for those with FOP due to surplus bone formation within the rib cage (Parker & Parker, 2007). The FOP median age of survival is approximately 40 years. Death often occurs due to complications of thoracic insufficiency syndrome (Kaplan et al., 2010).

In addition to breathing difficulties, some individuals are affected by limited range of motion of the neck. Moore et al. (2009) presented three case studies describing life-threatening deformity, chin-on-chest deformity. Chin-on-chest deformity occurs when the sternocleidomastoid muscle become ossified resulting in the restraint of the

head and face to the sternum or pectoral region. The case studies involved three girls ages nine months, seven years-old, and an eight year-old; all three had extreme stiffness and limited range of motion of the neck. Two of the three girls choose to undergo surgical intervention to correct the chin-on-chest deformity. As a result of the surgical intervention, both girls exhibited positive results. The third girl was unable to undergo surgical intervention due to safety measures. Typically surgery is not recommended because it can cause increased heterotropic ossification, however in these cases it was determined to be optimal.

Early diagnosis is essential, and unfortunately misdiagnosis of FOP is common because it is a rare genetic disorder and few doctors are aware of its classical symptoms. Misdiagnosis rates are approximately 80% or greater, which causes undo pain and suffering for individuals affected by FOP and their families (IFOPA, 2009g). FOP is commonly misdiagnosed as cancer, aggressive juvenile fibromatosis, or fibrous dysplasia (IFOPA, 2009g). Other common misdiagnosis include: isolated congenital malformations, brachydactyly, juvenile bunions, sarcoma, desmoids tumor, and lymphedema (Kaplan, Le Merrer, et al., 2008; Kaplan, Shen, et al., 2008).

When FOP is misdiagnosed, there is a lack of appropriate treatment. Various diagnostic procedures including bone scans, ultrasonic scans, open-surgery biopsies, and closed-needle biopsies all cause complications with individuals who have FOP (Kitterman, Kantanie, Rocke, & Kaplan, 2005). Diagnostic surgical interventions commonly worsen conditions and currently there are no effective treatments for FOP

(IFOPA, 2009b). In a case report completed by Zaghoul et al. (2008) a 10-week-old boy underwent a diagnostic biopsy and excision of a soft-tissue mass along the thoracolumbar spine. After the operation, the child had returned to the emergency room with discomfort and irritability. Due to the presenting factors of the surgical site, an examination of the lower extremities showed short great toes and malformations, thus resulting in a diagnosis of FOP. Within four weeks of the operation, heterotopic ossification had advanced along the entire surgical site. Early diagnosis of FOP is an important factor in the patients' safety because misdiagnosis may lead to harmful intervention and permanent complications.

Diagnosis of FOP can be made using contrast enhancement on CT and MRI scans to detect lesions before they become ossified. By diagnosing FOP early, further flare-ups can be avoided and clients can receive genetic counseling and optimal interdisciplinary team healthcare interventions (Smith, Athanasou, & Vipond, 1996). As well as helping with early diagnosing, PET/CT scans can show the progression of ossification and demonstrate other skeletal findings that are common for those diagnosed with FOP (Kulwin & Binkovitz, 2009). Additionally, other common symptoms of FOP to be considered include: large lateral masses on the cervical spine as well as small vertebral bodies, broad femoral necks, ossification at the insertion of the tendons on the long bones, and widened metaphyses (Smith, Athanasou, & Vipond, 1996). Awareness of the signs and symptoms of FOP in the early stages before the appearance of heterotopic

ossification can limit the disability and life-long harm of diagnostic errors and unnecessary procedures (Kaplan, Xu, et al., 2008).

Knowledge of the classical features of FOP and significance of the malformation of the big toe can aide in early diagnosis (Kaplan, Le Merrer, et al., 2008). In a study by Kaplan, Xu, et al. (2008), all seven children examined had congenital malformations of the big toes and were previously evaluated by orthopedic surgeons, but were not diagnosed until assessed by the study group for a possible diagnosis of FOP. Many clinicians are not aware of the classical symptoms of FOP, resulting in delay in diagnosis and healthcare interventions. There are no cures available for those with FOP (Kaplan, Le Merrer, et al., 2008; Kaplan, Xu, et al., 2008). However, treatment guidelines are available for those working with patients that have FOP.

Treatment guidelines for FOP are provided on the IFOPA website. These guidelines include: avoidance of deep tissue trauma such as intramuscular injections, protect the jaw, seek expert anesthesia professional, and consult expert doctors. These guidelines are suggested to avoid complications from trauma (IFOPA, 2009a). The authors Kaplan, Marine, et al. (2008) provide guidance points to help with the management of FOP. These guidelines as stated prior are provided for the following scenarios and topics: participation in activities, fall prevention, increased ossification (specifically to parts of the body), hearing, limb swelling, pulmonary function, education, and oral healthcare. Common suggestions for working with individuals with FOP include that joints should never be passively stretched, but that active range-of-motion exercises

are encouraged only if the movements are comfortable (Kaplan, Shore, & Pignolo, 2011; Kocyigit, Hizli, Memis, Sabah, & Memis, 2001).

Also, a home exercise program may be beneficial for those diagnosed with FOP (Ulusoy, 2010). Occupational therapists can play a role by educating the client on establishing a home exercise program that can help the client maintain current range of motion and address psychosocial issues that may arise. Additionally, OTs can collaborate with individuals diagnosed with FOP to increase their independence in dressing, toileting, bathing, mobility, education, work, home modifications, sexual activity, and other occupations the individual may hold. Furthermore, OTs should understand the individual's genetic makeup and predisposition conditions that can be a valuable aspect to intervention techniques. Occupational therapists understanding of the client's genetic makeup may help the individual with FOP increase or maintain their quality of life.

Role of Occupational Therapy

Occupational therapy can play a vital role in genetic research involving the care of individuals affected by FOP. Part of the role of occupational therapy is to help individuals be independent in their life. This may be in the form of lifestyle modification and environment changes. Occupational therapists are trained to understand the brain and body and to understand how the disease process may affect or be affected by the occupations or activities in which one may engage (Reynolds & Lou, 2009).

Occupational therapists are able to contribute to genetic research by engaging in new areas of clinical research and practice (Reynolds & Lou, 2009). For instance,

occupational therapists may conduct studies and contribute to research as well as prepare individuals for lifestyle changes they may face due to a diagnosis of a genetic condition such as FOP. Genetic research can be impacted by occupational therapists through: psychiatric genetic research, examining environmental influences on the progression of the disease, and monitoring the impact on function. Overall, occupational therapy being part of genetic research may consist of exploration of lifestyle and environmental experiences that influence the risk of psychiatric disorders and functional limitations that may be influenced by genetic disorders (Reynolds & Lou, 2009).

Additionally, OTs have inserted themselves into the field of genetic research. Occupational therapist's can provide valuable information and insight in regards to function in daily living activities for an individual newly diagnosed with a genetic disorder or over the progression of the disease. Occupational therapists should understand the client's genetic makeup and predisposition to conditions as it can be a valuable aspect to the treatment plan as well as how client factors, body structures, and functions may affect an individual diagnosed with a genetic disorder. Furthermore, understanding a client's genetic makeup may help OTs empathize with their clients. Currently, there are no effective remedies available for FOP, but an OTs understanding of genetics may help to promote clients to maintain or achieve a fulfilling life. The genetic information may allow OT's to understand and then elucidate how the disorder may effect a person's motivation, decision-making, and self-concept (Reynolds & Lou, 2009). Although OTs can participate in genetic research and educate those with FOP regarding their genetic

information, OTs can provide interventions regarding all activities of daily living (American Occupational Therapy Association [AOTA], 2008).

FOP results in a progressive loss of range of motion. Making activities of daily living more difficult. Occupational therapists can help individuals with this condition live a more fulfilling life (Kaplan, Le Merrer, et al., 2008). Other areas of occupation need to be addressed during treatment including: instrumental activities of daily living, work, education, social participation, leisure, rest and sleep, and play (AOTA, 2008). For those diagnosed with FOP, almost all strategies and interventions suggested should be compensatory, meaning that those with FOP should rely on equipment to help with function (Levy, Berner, & Bendixen, 2005).

Current treatment considerations as reported by Kaplan, Shore, & Pignolo (2011), consist of OT's addressing the following issues for those diagnosed with FOP; dressing, toileting, bathing, mobility, meal preparation, education, work, transportation, home modifications, and sexual activity. OTs can assist FOP clients with the occupation of dressing by recommending certain clothing and devices that will promote independence. Raised toilet seats or urinals may be utilized to increase functional performance in toileting. Also, OTs can recommend devices for personal hygiene. For the occupations of eating and meal preparation, the individual with FOP may benefit from the use of adapted utensils, tools, and environment modifications. Also, those with FOP may require their food to be pureed. Other occupational therapy interventions may address mobility issues

that an individual may be experiencing. Occupational therapists have the ability to recommend devices specific to mobility according to the individual's needs and wants.

The OT shall collaborate with other professionals in educational settings to address any needs the individual may have in the least restrictive environment. Transportation may be difficult for an individual with FOP. Therefore, OTs can recommend custom vans and adaptations to increase ease of access. In addition to transportation concerns, home modifications may be needed. Occupational therapists may need to complete a home evaluation to identify risks and problems that may need to change for the safety of the individual with FOP. The OT can recommend techniques to facilitate sleep participation and comfort of the individual (Kaplan, Shore, & Pignolo, 2011).

Also, OTs can address sexual activity with the individual. Certain positions may be difficult or painful for the client to perform. Occupational therapists can provide pillows or bolsters to help support the client in a more comfortable position. Occupational therapists can also encourage those individuals to discuss reproduction concerns with their physician (Kaplan, Shore, & Pignolo, 2011).

Because FOP is an inherited disease, concerns of passing the FOP mutation onto the clients offspring is a concern. If a parent has FOP his or her child will have a fifty percent chance of having the illness. Adding to the concern of pregnancy is the life threatening risks that are posed to both the mother and child. Various risks to the mother include the risk of flare-ups during pregnancy, breathing difficulties towards the end

stage of pregnancy, childbirth complications, general anesthesia risks for Caesarian delivery, and the risk of phlebitis and pulmonary embolism. Risks to the child is a fifty percent chance of inheriting FOP, risk of prematurity, risk of severe fetal distress, risk of cerebral palsy, and risk of complications from general anesthesia (Kaplan, F. S., Shore, E. M., & Pignolo, R. J., 2011).

Early Intervention

Stephans & Tauber, (2005) define intervention as the implementation of a program designed to maintain or enhance the development of a child in the natural environment and as a member of a family. Early intervention includes interventions designed “to prevent or minimize the physical, cognitive, emotional, and resource limitations of young children disadvantaged by biological or environmental risk factors” (Stephens & Tauber, 2005, p. 771). For those with FOP, OTs can design a home program that will allow the child to participate in occupations within their natural environment. Occupational therapy intervention includes educating the client and family members on specific safety precautions that need to be taken within their natural environment. Also, OTs should help those with FOP make adaptations in order to cope with changes in daily occupations. Through the use of educational techniques, OTs can help individuals with FOP increase their ability to cope with the illness. Additionally, OTs can provide education to those with FOP on techniques to minimize the progression rate of the disease. For example, prevention of pneumonia and decubitus ulcers for those who have FOP should be a major intervention technique (Hoeksma & Postuma, 1992).

Fall prevention

Fall prevention is an important factor OTs must consider when working with those diagnosed with FOP. According to Kaplan, Shore, & Pignolo (2011), falls for those with FOP may result in more severe injuries such as head injuries, loss of consciousness, concussions, and neck and back injuries due to decrease use of the upper extremities and anatomical abnormalities of the spine. Falls may cause severe injuries and ossification of the site injured by the fall. Because of this increased risk of injury, if the individual with FOP falls, immediate medical attention should be pursued for a complete evaluation and head or neck injuries should be considered serious until proven otherwise by their physician (Kaplan, Shore, & Pignolo, 2011).

Medication use, new environments, and medical conditions can inhibit proprioception of children, thus increasing their risk of falls (Cooper & Nolt, 2007). Caregivers need to be familiar with childhood development and milestones in order to be aware of new potential risks for falls in the home as failure to anticipate environmental hazards and risks for falls may lead to injury (McWilliams, 2011). An Infant/Child Home Fall Risk Tool has been developed to indicate a fall risk for patient's age's birth to 24 months and has been proven for practicality and easy interpretation of the results. The tool considers the following: crib/bed rails, safety restraint for changing tables, car seats, or bouncers, other children present, walkway clutter, supervision, head support, safety gates, co-sleeping of parent and child, and if the child is free from attached equipment (McWilliams, 2011). Occupational therapists can address the issues with falling by

recommending home modifications and increasing safety measures, mobility devices, and protective headgear (Kaplan, Shore, & Pignolo, 2011).

Education and Psychosocial Issues

When working with the parents of children diagnosed with FOP, in addition to emphasizing preventative measures, the OT must educate the parents on the importance of enabling leisure for their son or daughter. According to a study completed by Specht, King, Brown and Foris (2002), participation in leisure activities provides many positive benefits for individuals with or without a disability. These effects include increased self-esteem and self-worth, enjoyment, enhanced social interaction, and increased physical and mental health. Individuals with a disability have increased benefits from participation in leisure activities, and may develop a sense of belonging that can help give his or her life meaning (Specht, King, Brown, & Foris, 2002).

Parents of children diagnosed with chronic illnesses, such as FOP, experience an increase in stress, depression, health problems, and a lower quality of life. Parents of a child with a disability may have increased stress due to increased responsibilities and required demands including advocating for their child, managing medications, multiple appointments with professionals, managing nutrition, and coping with uncertainties about their child's illness (Strokes & Holsti, 2010). The increased stress has a direct impact on the socio-emotional and physical development of a child (Case-Smith, 2004).

A concept developed by sociologists, sense of coherence (SOC), can be used by occupational therapists as a framework for addressing parental stress. The sense of coherence is a measurement of a person's resilience and personal strength that gauges the individual's ability to respond to stressful situations. A low SOC is related to parental stress. Research has shown that this scale is a reliable, valid, and cross-culturally applicable tool to measure how people manage stressful life events (Strokes & Holsti, 2010).

As OTs working with the pediatric population, it is important to provide goals that are from a family-oriented perspective. This can help assure a more positive environment for the growth and development of pediatric clients (Strokes & Holsti, 2010). An additional concern regarding the influence of parents of children with a disability is that the parents have decreased social interaction which affects the entire family. Children learn how to interact with others by witnessing how their parents socialize (Case-Smith, 2004). Thus the parent-child relationship is an important aspect to address in FOP care plans.

The parent-child relationship is important during early development of any child and especially in young FOP patients. High quality parent-child interactions have been correlated to the child's ability to adapt to adversity, higher IQ scores, increased exploratory behavior, high-level sensorimotor development, and early language development. Occupational therapists can help support better parent-child interactions by including the parent within the intervention process. When parental stress is high, the

family interaction can be affected in a variety of ways including the quality of parent-child interactions, the child's experience within a larger environment not organized by their parents, and the health and safety provided by the family (Mayer, White, Ward, & Barnaby, 2002).

Individuals diagnosed with FOP will most often become more dependent on a caregiver as their disease progresses. There are a variety of early intervention approaches that are recommended for OTs to use when teaching caregivers. These include routines-based intervention, family-guided routines-based intervention, participation-based services, and learning opportunities. The key goal for routines- or activity-based approach is to promote a child's engagement in their family and community. Occupational therapists can teach caregivers to adapt the environment and use individualized learning strategies within family routines. The OT can teach the caregiver various interventions and encourage utilization of these interventions (Colyvas, Sawyer, & Campbell, 2010).

Assistive Technology

Supportive care is the primary focus for those with FOP. There are many available adaptations or modifications that OTs can prescribe for those with FOP (Kocyigit, Hizli, Memis, Sabah, & Memis, 2001). Shoes, canes, special toothbrushes, and wheelchairs are a few examples of available devices that can help a patient diagnosed with FOP achieve optimal independence in the home and community. In a research article by Hoeksma & Postuma (1992) an OT and a physiatrist designed a distinct standing and transport device

for a patient diagnosed with FOP who was experiencing difficulty with decubitus ulcers and mobility issues. The transport device was designed because the patient's feet had difficulty bearing his bodyweight during the day and he did not want to lie all day. The transport device consisted of front-wheel drive (to use on all terrains), the undercarriage consisted of a high-low control, and the frame was fitted to the patient's body, ROHO low-profile anti-decubitus mattress, a footboard, and driver controls adapted for the patient. Besides the transport device, other assistive devices were provided for the patient including an anti-decubitus mattress and rehabilitation shoes. The transport device allowed the patient to comfortably and independently go out into the community. In another article, (Levy, Berner, Sandhu, McCarty, & Denniston, 1999) a patient with advanced FOP was unable to stand upright or sit independently, was referred to a seating and positioning clinic. As part of the intervention process, the interdisciplinary team implemented the following devices: custom shoes and a customized power tilt wheelchair with custom foam. These devices were considered for the individual to allow community access and independent mobility within the home.

Other devices that an OT may use in the care of FOP patients include but are not limited to; sock donners, elastic shoelaces, long-handled shoehorns, Velcro closures, dressing sticks, zipper aids, raised toilet seats, custom-angled commodes, grab bars, long-handled sponges, modified reachers, shower benches, long-handled combs/brushes, electric toothbrush, water pics, adapted utensils, electrical appliances, hearing aids, and a special mattress (Levy, Berner, & Bendixen, 2005). Also, an OT may suggest ramps,

widened hallways, accessible kitchens, bathrooms, and bedrooms, reachable light switches, and control units for appliances, doors, televisions, and telephones.

Aquatics Therapy

Aquatics may be utilized as a beneficial occupational therapy intervention by instilling meaning and purpose for the client diagnosed with FOP by allowing participation in desired occupations. Occupational therapists may use water based therapy as a way to focus intervention on performance skills or physical aspects of the client as well as emotional and social participation aspects. The aquatic environment may allow the client to perform occupation based activities that they may have difficulty performing on land. This may be possible due to the water's buoyancy that reduces the effects of gravity and the water's warm temperature may provide relaxation for clients that experience stress, pain, and/or muscle tightness.

Overall, benefits of aquatic therapy may include improved psychosocial factors such as interests, social conduct, self-esteem, and trust. Also, cognitive skills may be addressed including attention span, sequencing, and memory skills. Specifically occupational intervention through aquatics may address goals that consist of health promotion, leisure activities, social participation, pain management, self-exploration, and relaxation techniques. (Fischer, et al., 2001)

According to Kaplan, Shore, & Pignolo (2011), aquatic therapy may be utilized with individuals diagnosed with FOP to perform active range of motion, cardiopulmonary

and resistive exercise in a safe-low impact environment. In order to enter and exit the pool, individuals with FOP may need to utilize lifts, elevators, or ramps.

Overall, FOP has can have an effect on all areas of occupation. Occupational therapists play an important role in healthcare for individuals who have FOP. Occupational therapists can advocate for clients with FOP by helping other professions recognize the signs and symptoms of the disease. Occupational therapists can also assist in educating clients on safety precautions to avoid falls as well as make various recommendations for equipment to assist in performance of ADL's.

Summary

Fibrodysplasia Ossificans Progressiva is a rare and disabling genetic disorder that results in ossification of the soft tissues (Kaplan, Xu, et al., 2008; Kocyigit, Hizil, Hemis, Sabah, & Memis, 2001). There is a lack of research and literature on therapeutic intervention for those with a diagnosis of FOP. However, OTs can help those with FOP increase their quality of life through enhancement of activities of daily living (Kaplan, Le Merrer, et al., 2008). Occupational therapy can help those diagnosed with FOP increase their functional independence throughout their life-span and provide supportive care.

The next chapter is the methodology chapter that includes information on the development process and instruments used to collect data, to complete this scholarly project.

CHAPTER III

METHODOLOGY

The development process of this scholarly project began upon the authors' inquiry of FOP. The authors learned about an individual diagnosed with this disease and realized the impact FOP has on an individual's ability to function. This led the authors to research what evidence based occupational therapy literature available related to FOP. However, there were few evidence based articles and the authors determined there was a need for further exploration of literature related to occupational therapy and FOP.

After recognizing the need, the authors decided it was important to develop a manual for OTs working with individuals diagnosed with FOP. A literature review was completed to determine what FOP is, the severity of the disease, and what research was available for OTs regarding FOP. This led to a further review of current research regarding the role of OTs with an individual diagnosed with a rare disease. The authors located research articles by using the following terminology: "Fibrodysplasia Ossificans Progressiva", "FOP", and "heterotrophic ossification".

After reviewing the current research related to FOP and occupational therapy, the authors interviewed an individual afflicted with FOP. The authors discovered that the individual's perspective was beneficial in the development of the manual. Providing OTs with this manual would give them important guidelines to consider when providing

therapy to individual's with FOP. The authors also contacted Dr. Kaplan, an expert in the field of FOP, for his expertise regarding his work with FOP and his suggested treatment guidelines.

The development of the manual was based upon the current research literature regarding OTs role in providing care for individuals diagnosed with FOP. This manual was developed for all OTs providing care to individuals diagnosed with FOP. Through implementation of this manual, it is anticipated that more OTs will feel they have resources available to allow them to provide quality care to those diagnosed with FOP.

The next section is the occupational therapy manual that includes information on the strategies and methods to be used during intervention with individuals diagnosed with FOP.

CHAPTER IV

PRODUCT

This chapter consists of the manual to help guide treatment for an occupational therapist working with an individual with FOP. This manual begins with a synopsis of symptoms related to FOP because many individuals are not aware of this disorder. The authors then provide guidelines to follow, strategies for building rapport, and the Model of Human Occupation (MOHO) that guides the entire manual. The authors then recommend various MOHO assessments that can be used when working with this population.

This manual also includes important issues related to caring for individuals with FOP. These include psychosocial issues, information for caregivers, early intervention strategies, falls prevention, and aquatic therapy. These areas of the manual include general guidelines of what the occupational therapist or caregiver can focus on to optimize treatment.

CARING FOR INDIVIDUALS WITH FIBRODYSPLASIA OSSIFICANS PROGRESSIVA (FOP): A TREATMENT MANUAL FOR OCCUPATIONAL THERAPISTS



Printed with permission from K. Fenton

Amanda Rositas, MOTS

Kathryn Zavoral, MOTS

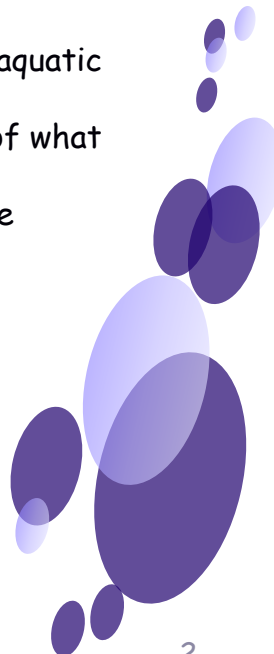
Mandy Meyer, Ph.D.




INTRO TO MANUAL

The following is a manual to help guide treatment for an occupational therapist working with an individual with Fibrodysplasia Ossificans Progressiva (FOP). This manual begins with a synopsis of signs and symptoms of FOP because many individuals are not aware of this disorder. The authors then provide guidelines to follow, strategies for building rapport, and the Model of Human Occupation (MOHO) that guides the entire manual. The authors then recommend various MOHO assessments that can be used when working with this population.

This manual also includes important issues related to caring for individuals with FOP. These include psychosocial issues, information for caregivers, early intervention strategies, falls prevention, and aquatic therapy. These areas of the manual include general guidelines of what the occupational therapist or caregiver can focus on to optimize treatment.





In addition, the authors addressed activities of daily living, instrumental activities of daily living, rest and sleep, school, and home modifications.

These areas will also refer to assistive technology that can help facilitate participation in the individual's occupations.

Lastly the occupational therapist's role in genetic research is included in this manual. This is important because the occupational therapist can help educate the client and client's family regarding genetic information as well as conduct research studies.



Table of Contents

What is FOP_____	5
Occupational Therapy Intervention Guidelines_____	8
Building Rapport with Individuals diagnosed with FOP_____	10
The Model of Human Occupation_____	15
Assessments_____	21
Psychosocial Issues_____	32
Caregivers_____	34
Early Intervention_____	38
Falls Prevention_____	40
Aquatic Therapy_____	43
Activities of Daily Living_____	47
Instrumental Activities of Daily Living_____	54
Rest and Sleep_____	57
School_____	59
Home Modifications_____	61
Occupational Therapist's Role in Genetics_____	64
References_____	66



What is FOP?

"The doctor said, 'We don't know how long she will live, we don't know anything...we will be lucky if she makes it past the age of six.'

(K. Fenton, personal communication, November 7, 2011)

"Every case isn't the same.. Everybody is unique."

(K. Fenton, personal communication, November 7, 2011)



FOP

Genetic disorder in which muscle tissue and connective tissue are progressively replaced by bone

- Disease produces bone outside skeleton
- Affects 1 in 2 million people
- 700 confirmed cases around the world
- Gene mutation
 - Autosomal dominant pattern
 - May result from new mutation
- *ACVR1* gene
 - Always activated and causes overgrowth of bone and fusion of joints
- Definitive genetic testing available to confirm diagnosis

(Parker & Parker, 2007; Kaplan, Le Merrer, et al., 2008, IFOPA 2009b)

"You know that saying, 1 in a million? I am 1 in 2 million."

(K. Fenton, personal communication, November 7, 2011)

"They said it could be cancer."

(K. Fenton, personal communication, November 7, 2011)

Common Symptoms/Characteristics

- Short, malformed great toes



Printed with permission from K. Fenton

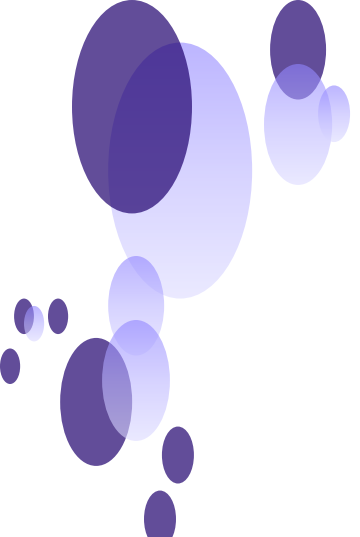
"My mom started noticing grapefruit size nodules where my glands were."

(K. Fenton, personal communication, November 7, 2011)


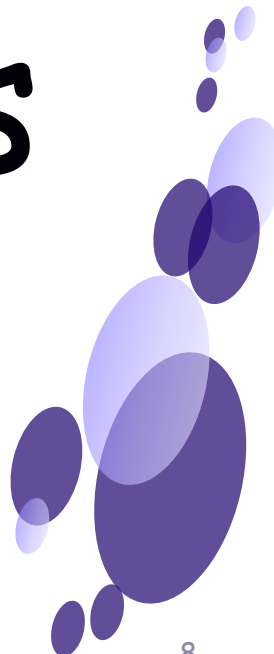
- Fibrous masses in soft connective tissue
 - Turn into mature bone
- Extra bone formation starts in childhood or later in life
- Often advances in patterns
 - Proximal to distal
 - Upper extremities to lower extremities
 - Axial to appendicular
- Scalp nodules
- Restricted mobility
- Progressive loss of range of motion
(Piram et al., 2011, Cohen et al., 1993, IFOPA, 2009d)

"I can't move my head, I've never been able to... I had to turn my whole body."

(K. Fenton, personal communication, November 7, 2011)



Occupational Therapy Intervention Guidelines





Occupational Therapy Intervention Guidelines

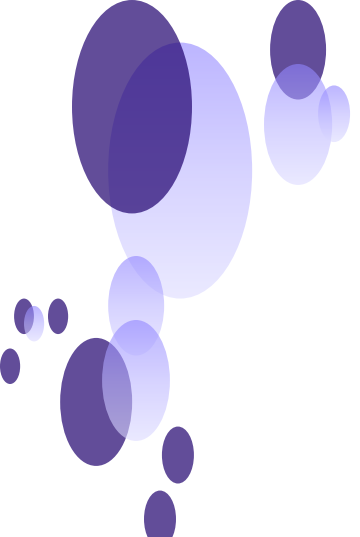
Collaborate with client's physician before and during treatment if questions arise.

Provide insight over the progression of the disease and how the individual's functional ability will be affected in regards to desired occupations.


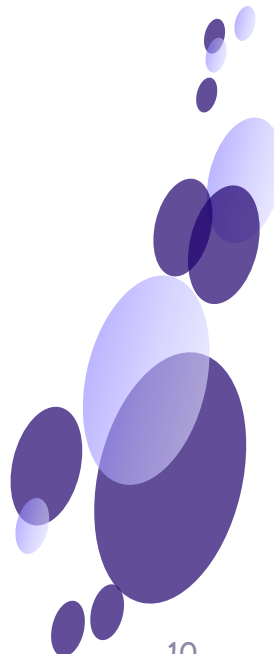
- Understand how client factors and body structures will be affected
- Understand that FOP may effect the individual's motivation, decision making, and self-concept


(Reynolds & Lou, 2009)

- Almost all of the strategies and interventions should be compensatory
- Avoid hands-on techniques
- Make referrals for pain management as needed



Building
Rapport with
Individuals
diagnosed with
FOP






When a therapist is working with a client with FOP building rapport is very important. This should be done before any other assessments and throughout the treatment process. Rapport is important because it helps establish a good therapeutic relationship between the client and therapist. When a therapeutic relationship is established, trust, and willingness for client to be successful in the treatment program is more common.

Building rapport is defined as deliberately making the client feel at ease. It is important to establish a routine initially and communication style that makes the client feel most comfortable.

(Taylor, R. 2008)



Examples of rapport building techniques include the following:

- Maintain appropriate eye contact and greet in a friendly manner
- Asking the client how he/she would like to be addressed
- Introducing yourself with or without a formal prefix (depending on your best judgment). With children it is suggested to use a formal prefix followed by a first name
- Ask the client if you are pronouncing his/her name correctly
- Orienting the client to the environment and setting
- Making small talk about relatable topics
- Asking how the day is going or how he or she is feeling
- Letting the client know that you know something about them (if appropriate)
- Share facts about yourself


(Taylor, R. 2008)





The Model of Human Occupation





The Model of Human Occupation (MOHO) will be implemented throughout this occupational therapy manual. The Model of Human Occupation will be used throughout the occupational therapy intervention process. This model is a client-centered based model that is appropriate for this manual.

For individuals with FOP, the therapeutic interventions are strategically implemented according to their characteristics and the ways the individual behaves, thinks, and feels during completion of their occupations.

According to MOHO, humans are thought of as having interrelated aspects; volition, habituation, and performance capacity (Kielhofner, 2008).



Volition

Volition is referred to as the motivation for completing occupations.

- personal capacity and effectiveness
- self-worth that is attached to what the individual does
- satisfaction experienced when doing occupations

(Kielhofner, 2008; Cole & Tufano; Kielhofner, Forsyth, Kramer, Melton, & Dobson, 2009).

An individual with FOP will anticipate, choose, experience, and interpret what he or she does through thoughts of effectiveness, self-worth, and satisfaction of the occupation.




Habituation

Habituation is referred to the patterns of an individual's behavior in relation to:

- Temporal environment
- Physical environment
- Social environments

(Kielhofner, 2008; Cole & Tufano, 2008; Kielhofner et al., 2009).

For someone with FOP, a habit may be the repeated action of transferring in and out of their wheelchair to their bed (repeated action that establishes a pattern) in their bedroom (consistent environment). Also, an individual with FOP may identify themselves with the role of a student, a role that is associated to a social status. As a student an individual will partake in patterns of behaviors that reflect their role as a student.





Performance Capacity

Performance capacity refers to bodily systems and mental/cognitive abilities that an individual uses when participating in occupations (Kielhofner, 2008; Cole & Tufano, 2008; Kielhofner., 2009).

Bodily structures to consider when providing therapy to individuals with FOP include:

- Musculoskeletal
- Neurological
- Cardiopulmonary

It is important to be aware of the client's experiences of having limitations in performing occupations and especially the limitations of performance.



Environment

Environment consists of:

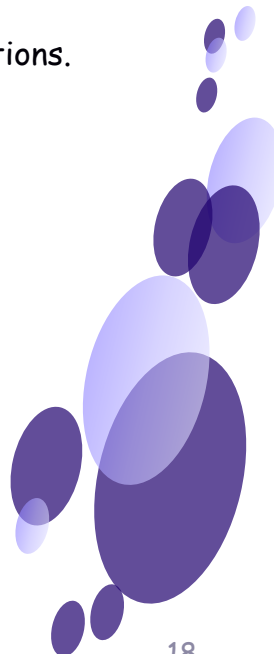
- Physical Context
- Sociocultural context

The environment incorporates:

- The space an individual occupies
- The objects the individuals use
- Interactions with other individuals

(Kielhofner, 2008).

For individuals with FOP, it is essential that the occupational therapist considers all supports and barriers the environment has on the individual being successful in participation of occupations.






Six Steps of Therapeutic Reasoning

During evaluations, the occupational therapist should implement the following six steps:

- *Generating questions*
- *Collecting information about the client*
- *Use information gathered to explain the client's situation*
- *Generate goals and strategies for therapy*
- *Implement and monitor therapy*
- *Determine therapy outcomes*

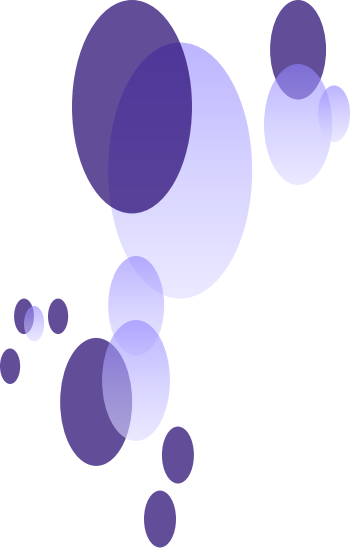
(Kielhofner, 2008).



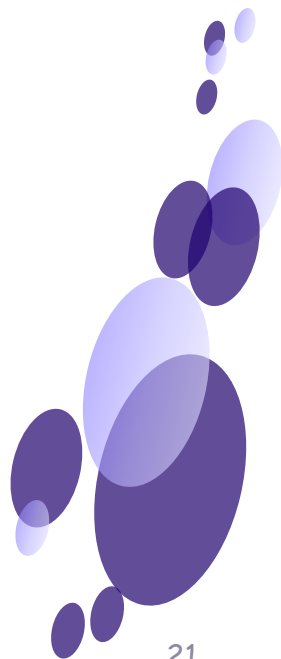
During the therapy process, clients can contribute to their change through occupational engagement by:


- Choosing, or making decisions, committing, exploring, identifying, negotiating, planning, practicing, reexamining, and sustaining their engagement in desired occupations (Kielhofner, 2008; Kielhofner et al., 2009).

Overall, occupational therapists are able to implement MOHO throughout the therapy process to promote client-centeredness and successful occupational engagement for the clients. The Model of Human Occupation allows occupational therapists to think of clients holistically as well as implement occupation-based activities. The Model of Human Occupation considers individuals to be composed of three interrelated aspects; volition, habituation, and performance capacity (Kielhofner et al., 2009).



Assessments





The following are various assessments that are applicable for therapists to use when working with clients who have FOP. A clinician should use his or her best judgment when deciding which assessments are appropriate. This information can be utilized as helpful suggestions, but one should not be limited to only these options.



Active Range of Motion

•**Author:** Hazel M. Clarkson, M.A., B.P.T.

•**Format:** Observation of multiple joints simultaneously, one at a time, and/or bilaterally

•**Purpose:** Provides information about the individual's willingness to move, coordination, level of consciousness, attention span, joint ROM, movements that cause or increase pain, muscle strength, ability to follow directions, and performance in functional activities

Because FOP affects range of motion throughout all joints, individuals would benefit from having a Functional Range of Motion assessment performed by a therapist.

This is beneficial because the therapist can monitor the client's progress as far as maintaining range of motion for as long as possible.

- Only active range of motion should be tested
- Avoid passive range of motion

•**Population:** All ages

•**Time Required:** Varies

(Clarkson, 2000)



Model of Human Occupation Screening Tool (MOHOST version 2.0)

- Authors:** Sue Parkinson, DipCOT; Kirsty Forsyth, PhD, SROT, OTR; Gary Kielhofner, DrPH, OTR, FAOTA
- Format:** Observation, discussion, and record review
- Purpose:** Screening tool to determine the need for occupational therapy services and to document occupational functioning using the concepts of the Model of Human Occupation
- Population:** Clients who have mental health problems and trouble tolerating lengthy interviews
- Time Required:** Observations can last a few days to a few weeks. Scoring lasts 10-20 minutes


(Schultz-Krohn, 2007)



Child Occupational Self Assessment (COSA version 2.1)

- Authors:** Kathi Baron, MS, OTR; Gary Kielhofner, DrPH, OTR, FAOTA; Anita Iyengar, MS, OTR; Victoria Goldhammer, OTS; Julie Wolenski, OTS
- Format:** Self-report questionnaire; administered individually or in a small group
- Purpose:** Assessment and outcome measure based on MOHO, it is used to collect data on the individual's self-perception of occupational performance
- Population:** ages 8-13
- Time Required:** 10-20 minutes

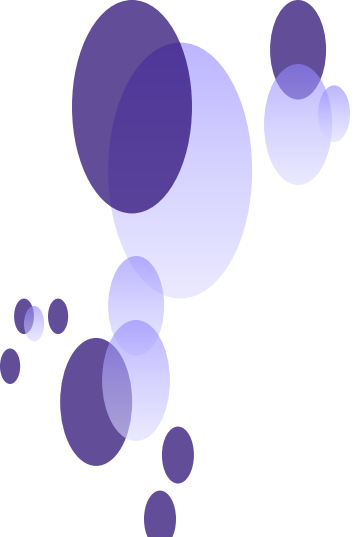
(Schultz-Krohn, 2007)



Pediatric Volitional Questionnaire (PVQ version 2.0)

- Authors:** Carmen Gloria de las Heras, MS, OTR/L; Rebecca Geist MS, OTR/L; Gary Keilhofner DrPH, OTR, FAOTA; Yanling Li, MA; Semonti Basu MS, OTR; Ana Kafkes MS, OTR; Rebecca Geist, MS, OTR/L; Gary Keilhofner DrPH, OTR, FAOTA
- Format:** Observation-based behavior rating scale
- Purpose:** Based on MOHO, this assessment is appropriate for individuals who cannot express themselves verbally
- Population:** children 2-7 years of age
- Time Required:** 10-30 minutes, depending on child's tolerance

(Schultz-Krohn, 2007)



Occupational Therapy Psychosocial Assessment of Learning (OT PAL), Version 2.0

- Authors:** Sally Townsend, Paula Carey, Nancy Hollins, Christine Helfrich, Melinda Blondis, Amanda Hoffman, Lara Collins, Julie Knudson, Angela Blackwell
- Format:** Observation and descriptive assessment tool
- Purpose:** This instrument focuses on psychosocial skills and the fit between the child and school environment
- Population:** 6-12 years old
- Time Required:** Varies

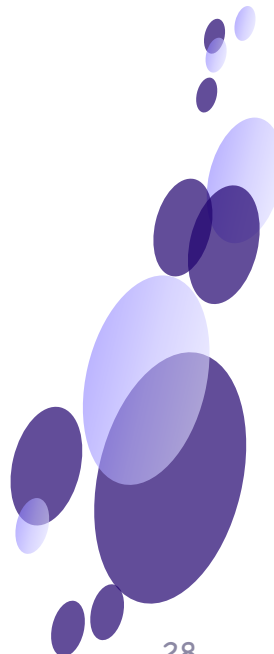
(Martin, 2007)



The Activities of Daily Living (ADL) Index

- **Authors:** Kazim A. Sheikh, Dennis S. Smith, Thomas W. Meade, E. Goldenberg, P.J. Brennan, Glynda Kinsella
- **Format:** Interview and observation-based rating scale
- **Purpose:** Assess a person's ability to perform basic ADL's and mobility tasks associated with ADL's
- **Population:** People with chronic disability
- **Time Required:** 20 minutes

(Furphy, 2007)





Matching Person and Technology (MPT)

Clients with FOP will often require assistive technology to aid with their mobility and ADL's.

- **Authors:** Marcia J. Scherer
- **Format:** Self-report checklist and questionnaire
- **Purpose:** Evaluating and selecting assistive technologies to promote independence in the client's performance at home, school, workplace, and health care settings
- **Population:** Clients, students, or employees with physical or sensory disabilities; reading ability is required
- **Time Required:** 15 minutes per form

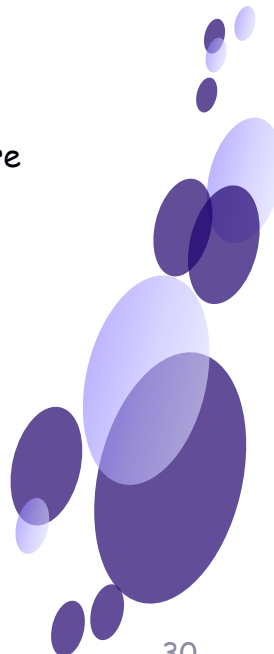
(Furphy, 2007)



Children's Assessment of Participation and Enjoyment (CAPE)

- **Authors:** Gillian King, Mary Law, Susanne King, Patricia Hurley, Peter Rosenbaum, Steven Hanna, Marilyn Kertoy, Nancy Young
- **Format:** Picture-based questionnaire and rating scale, administered by caretaker or evaluator
- **Purpose:** Measure a child's participation in, enjoyment of, and preferences for activities other than school activities
- **Population:** 6 to 21 years of age, with or without disabilities, who are able to discriminate between different activities
- **Time Required:** 30-45 minutes to administer and score

(Martin, 2007)





Infant/Child Home Fall Risk Tool

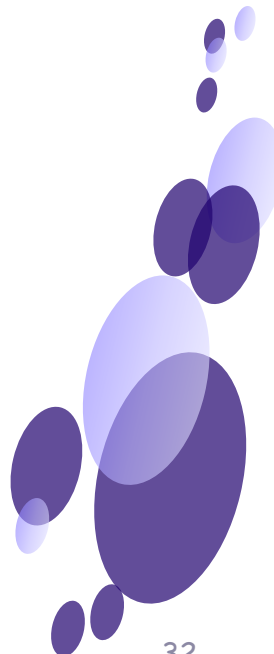
- **Authors:** James R. McWilliams
- **Format:** Questionnaire
- **Purpose:** Indicate the fall risk for infants and children ages birth to 24 months
- **Population:** Infants and toddlers
- **Time Requirement:** Varies
- **Considerations/Assess:**
 - Crib/bed rails
 - Safety restraints
 - Changing tables
 - Car seats
 - Bouncers
 - Other children present
 - Walkway clutter
 - Supervision
 - Head support
 - Safety gates
 - Co-sleeping of parent and child
 - Child free from attached equipment



"I talk(ed) to
my FOP
friends."

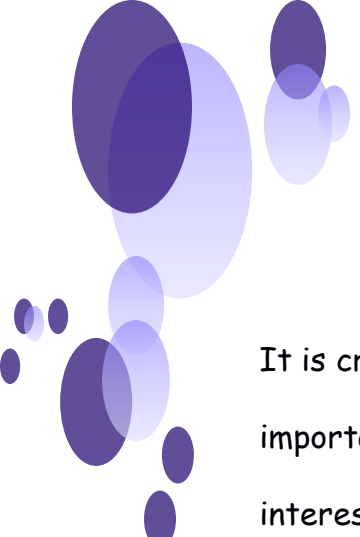
(K. Fenton, personal
communication, November 7,
2011)

Psychosocial Issues



"In 10th grade, I
did 'Make a Wish'
my wish was for us
to go on a cruise...
all six of us."

(K. Fenton, personal communication,
November 7, 2011)



It is crucial to educate the clients and caregivers on the importance of maintaining and seeking out leisure activities and interests.

Leisure activities may:

- Develop a sense of belonging
- Increase self-esteem and self-worth
- Enhance enjoyment
- Enhance social interaction
- Improve physical and mental health

It is also vital to educate clients, family members, and caregivers that the individual with FOP may experience:

- Increased stress
- Depression
- Health problems
- Lower quality of life

Therapist should also educate the clients, family members, and caregivers on ways to cope and deal with these difficulties.

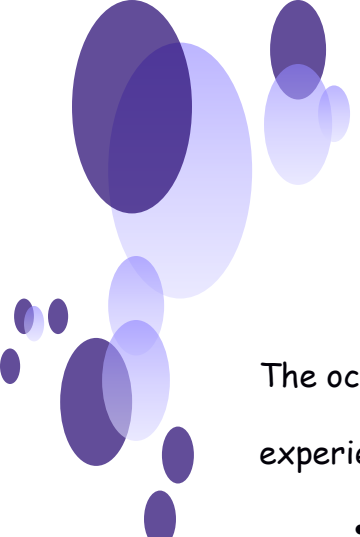
(Case-Smith, 2004; Specht, King, Brown, & Foris, 2002; Strokes & Holsti, 2010)



Caregivers

"After I was diagnosed, my mom walked around the nurses station crying.. My mom thought it was her fault."


(K. Fenton, personal communication, November 7, 2011)



The occupational therapist must be aware that caregivers may experience:

- Increased stress due to increasing responsibilities
- Increased demands for advocating for their child
- Increased demands of managing the child's medications
- Increased demands of managing multiple appointments with numerous professionals
- Increased demands of managing child's nutritional needs
- Difficulties in coping with uncertainties of the child's diagnosis

(Case-Smith, 2004; Specht, King, Brown, & Foris, 2002; Strokes & Holsti, 2010)



Occupational therapy goals should take into consideration all family members to ensure a positive environment for growth and development.

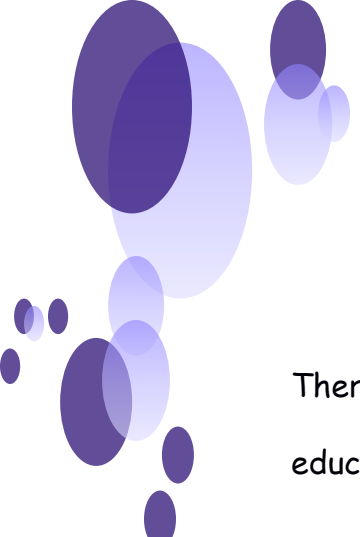
Occupational therapists should consider the importance of the child's social interaction with:

- Caregivers
- All family members

Occupational therapists should support better caregiver-child interactions:

- Include caregiver and siblings during the intervention process

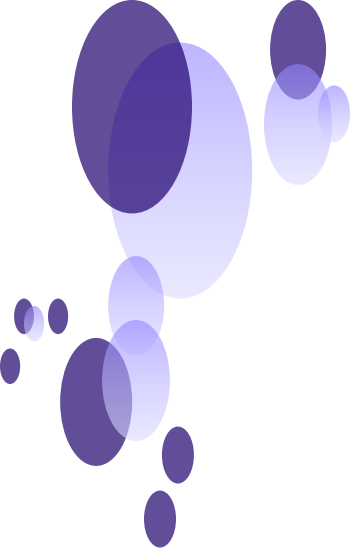
(Colyvas, Sawyer, & Campbell, 2010; Mayer, White, Ward, & Barnaby, 2002)



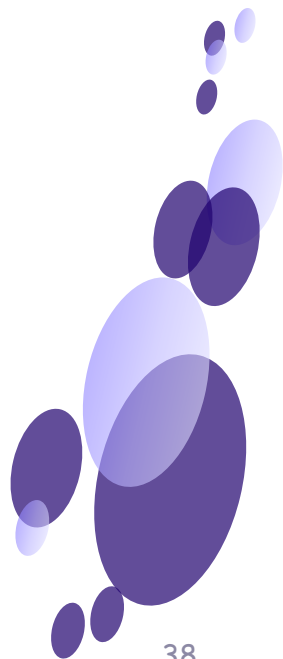
There are many interventions that may be utilized to educate caregivers;


- Key goal -- to promote the child's engagement in their family and community
 - Routine-based interventions
 - Family-guided routine based intervention
 - Participation based services
 - Learning opportunities
 - Techniques to adapt the environment
 - Individualized learning strategies for family routines

(Colyvas, Sawyer, & Campbell, 2010; Mayer, White, Ward, & Barnaby, 2002)



Early Intervention





Occupational therapists must assist in designing a home program for individuals with FOP.

- To allow the individual to participate in occupations within their natural environment


Therapists should help individuals cope with lifestyle changes and cope with diagnosis of FOP and provide education on techniques used to minimize the progression rate of the disease:

- avoid ulcers
- avoid contracting pneumonia or influenza
- avoid falls
- avoid trauma
- Protect head



Fall Prevention

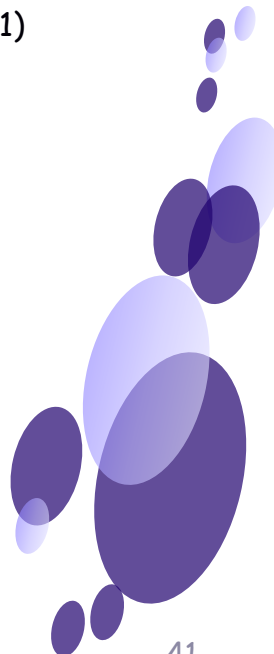




Fall prevention is essential when providing therapy for clients with FOP because any trauma or injury may cause further progression of the disease.

- Educate the client and caregivers regarding the side effects of medication
- Educate the client and caregivers on environmental factors that may cause or increase the chances of falls
- Educate client and caregivers to anticipate hazards
- Utilize the Infant/Child Home Fall Risk Tool to indicate fall risks for the individual
 - Used as an education tool for the parents and caregivers

(Kaplan, Shore, & Pignolo, 2011; McWilliams, 2011)



- Recommend home modifications
- Increase safety measures
- Recommend protective headgear



Printed with permission by Patterson Medical, 2011

"I had to wear helmets because I would fall a lot."

(K. Fenton, personal communication, November 7, 2011)


- Educate client, family members, and caregivers on the importance of seeking medical attention from their physician immediately after the individual with FOP falls

(Kaplan, Shore, & Pignolo, 2011; McWilliams, 2011)



Aquatic Therapy





Occupational therapists should gain certification in aquatic therapy.

Aquatic therapy offers a safe and low impact environment:

- Warm water to facilitate pain relief
- Allows individuals with FOP to perform active range of motion
- Cardiopulmonary exercise
- Resistive exercise
- Modified lifts
- Elevators
- Ramps

Therapists should address goals such as health promotion, leisure activities, social participation, pain management, self exploration, and relaxation techniques

Almost all of the strategies and interventions should be compensatory.

(Aquatic Exercise Association, 2010; Fisher, et al., 2001; Kaplan, Shore, & Pignolo, 2011)



Aquatic Therapy considerations:

- Class format
 - Warm-up
 - Conditioning phase
 - Cool down
- Water quality maintained
- Water temperature
 - Depends on child's age and activity involved
- Water depth
- Pool entry and exit
- Air temperature and humidity
- Air quality
- Intensity
 - Bouyancy, resistance
- Music

"I would use the whirlpool twice a week for aches and pains."

(K. Fenton, personal communication, November 7, 2011)



Aquatic Therapy considerations continued:

- Arm positioning
- Equipment utilized
 - Flotation devices
- Proper footwear
- Proper clothing
 - Professional
 - Individual with FOP
- Hydration
 - Professional and Individual with FOP
 - Drink water before, during, and after
- Safety instruction for other professionals, clients, family members, and caregivers



Activities of Daily Living

Eating:

- Full eating evaluation should be considered
- Occupational therapist has to be certified in this area



Printed with permission from K. Fenton



Printed with permission by
Patterson Medical, 2011



Printed with permission from K. Fenton

Dressing:

- Pull over shirts and blouses
- Zipper aids
- Elastic waistbands for pants
- Velcro closures
- Elastic shoe laces
- Sock donners
- Long handled shoe horns and reachers



Printed with permission by
Patterson Medical, 2011



Printed with permission by
Patterson Medical, 2011

Bowel and Bladder management:

- Raised toilet seats
- Custom-angled commodes
- Beside urinals



Printed with permission by
Patterson Medical, 2011



Printed with permission by
Patterson Medical, 2011

"My bottom teeth were turning black because I couldn't get between the teeth. The bottoms were all decaying (teeth).. They took them all out completely."

(K. Fenton, personal communication, November 7, 2011)

Personal Hygiene:

- Long handled sponges
- Long handled combs
- Modified reachers
- Electric toothbrushes
- Water pics
- Suction devices
- Shower chair or shower gurney



Printed with permission by
Patterson Medical, 2011



Printed with permission by
Patterson Medical, 2011



Printed with permission by
Patterson Medical, 2011

Functional Mobility (depends on the disease progression stage):

Transfers in and out of bed, chair, car, tub, toilet, floor, tub/shower, etc

- Canes
- Walkers
- Crutches
- Custom shoes
- Standing device or transport device
- Power wheelchairs
 - Customized seating
 - Power seat elevation and depression
 - Anterior and posterior tilt and recline function
 - Lap trays with mounts

'How much is your wheelchair?' ... "I'm like, it's my car!"

(K. Fenton, personal communication, November 7, 2011)



Printed with permission from K. Fenton



Sexual Activity:


- Requires thorough consideration
- Recommend the individual with FOP to discuss sexuality and reproduction with their physician
- Pillows
- Bolsters
- Postures

(Kaplan, Shore, & Pignolo, 2011)



Instrumental Activities of Daily Living





Almost all of the strategies and interventions should be compensatory.

Meal Preparation:

- Electrical can and jar openers
- Cutting boards with spikes to hold food while preparing
- Food may be pureed

(Kaplan, Shore, & Pignolo, 2011)

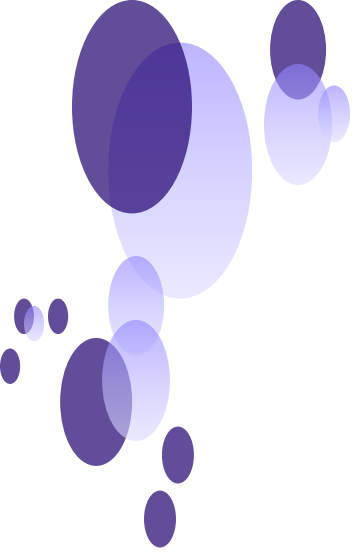


Transportation:

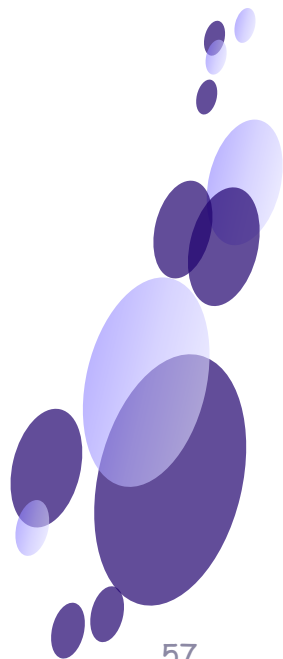
- Vans can be customized for the individual with FOP
 - Ramps
 - Lifts
 - Roofs can be raised
 - Floors can be lowered
 - New controls and motors
 - to lower ground clearance

(Kaplan, Shore, & Pignolo, 2011)





Rest and Sleep



Almost all of the strategies and interventions should be compensatory.

Therapists should facilitate sleep participation and comfort for the individual with FOP:

Tilt Beds

- From vertical to horizontal and vice versa
Redistribute pressure, provide comfort
and protect skin integrity
- Specialized Mattresses
- Overlays

(Kaplan, Shore, & Pignolo, 2011)

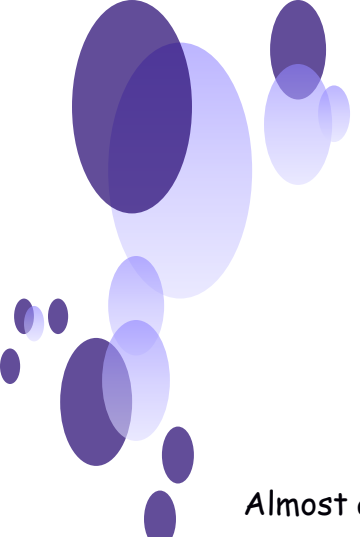


Printed with permission from K. Fenton



School





Almost all of the strategies and interventions should be compensatory.

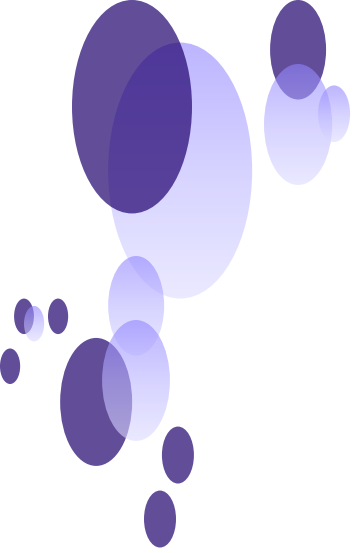
Occupational therapists should encourage an Individualized Education Plan (IEP) including:

- Occupational Therapy
- Physical Therapy
- Speech Therapy

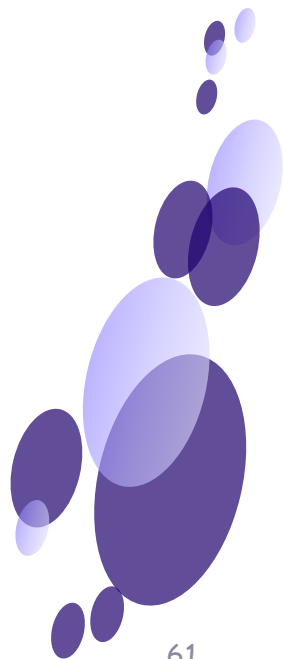
Additionally, the plan should:


- Encourage intellectual pursuits
- Encourage computer skills
- May need to make referrals
- Vocational Rehabilitation, to help those with FOP enter and/or remain in the work force

(Kaplan, Shore, & Pignolo, 2011)



Home Modifications





Home evaluation should be completed to determine risks and problems that need to be addressed to ensure safety of the individual with FOP

Almost all of the strategies and interventions should be compensatory:

- Widened doorways and hallways
- Installation of grab bars
 - Bathrooms
 - Hallways
- Rotating shelves
- Use of stools or elevated platforms
- Eliminate or minimize steps
- Accessible rooms
 - Bathroom
 - Kitchen
 - Bedroom
 - Living room



Printed with permission by
Patterson Medical, 2011

- Install environment control units to operate:
 - Appliances
 - Doors
 - Televisions
 - Telephones
- Reachable light switches
(Kaplan, Shore, & Pignolo, 2011)

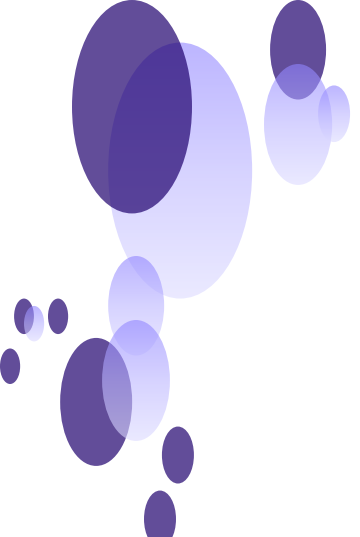


Printed with permission by Patterson Medical, 2011



Occupational Therapist's Role in Genetics





Occupational therapist should prepare affected individuals for lifestyle changes related to their diagnosis of a genetic condition such as FOP:

- Provide valuable information for individuals newly diagnosed with FOP
- Educate individuals and their family members and caregivers regarding their genetic information

Occupational therapists can engage in clinical research and practice related to FOP:

- Conduct studies and contribute to research


Therapists should also understand the genetic makeup of FOP clients to increase empathy.

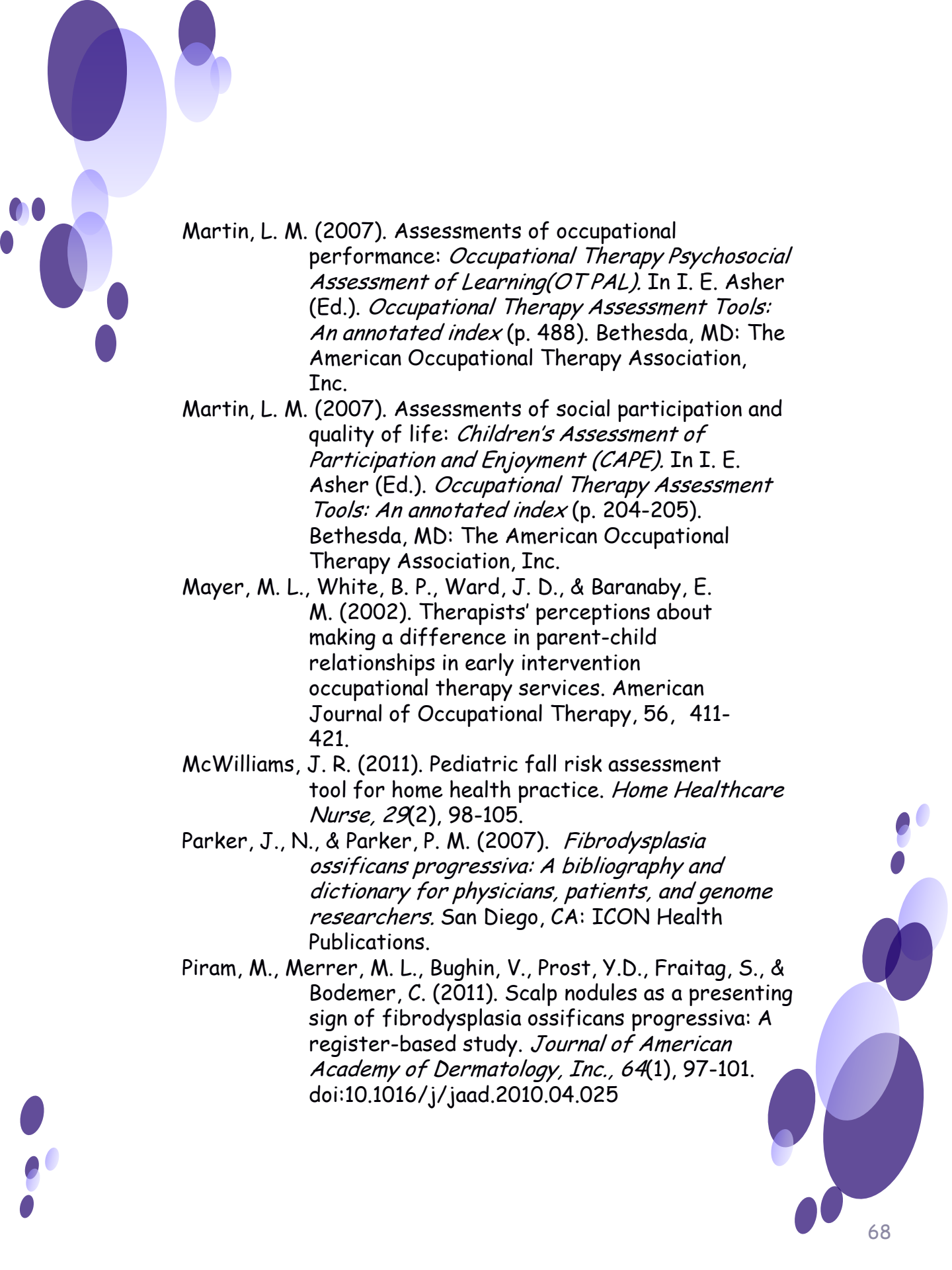
(Reynolds & Lou, 2009)



References

- Aquatic Exercise Association. (2010). Standards and Guidelines. Retrieved from <http://www.aeawave.com>
- Case-Smith, J. (2004). Parenting a child with a chronic medical condition. *American Journal of Occupational Therapy, 58*, 551-560.
- Clarkson, H. M. (2002). *Musculoskeletal assessment: Joint range of motion and manual muscle strength*. Baltimore, MD: Lippincott Williams & Wilkins.
- Cohen, R. B., Hahn, G. V., Tabas, J. A., Peepers, J., Levitz, C. L., Sando, A., ... Kaplan, F. (1993). The natural history of heterotopic ossification in patients who have fibrodysplasia ossificans progressiva. *The Journal of Bone and Joint Surgery, 75*(2), 215-219.
- Cole, M. B. & Tufano, R. (2008). Model of human occupation. *Applied theories in occupational therapy: A practical approach* (pp. 95-104). Thorofare, New Jersey: SLACK Incorporated.
- Colyvas, Sawyer, & Campbell. (2010). Identifying strategies early intervention occupational therapists use to teach caregivers. *American Journal of Occupational Therapy, 64*(5), 776-785.
- Fischer, M. S., Kratz, A., Jimenez, B., Watson, C., Spence, C., Sanford, T., Goertz, H., & Scolaro, M. (2001). Aquatic therapy: An occupational perspective. *OT Practice Online, 5*, 14-16.
- Furphy, K. A. (2007). Assessments of activities of daily living and instrumental activities of daily living: *Activities of Daily Living Index*. In I. E. Asher (Ed.). *Occupational Therapy Assessment Tools: An annotated index* (p. 57). Bethesda, MD: The American Occupational Therapy Association, Inc.

- 
- Furphy, K. A. (2007). Assessments of activities of daily living and instrumental activities of daily living: *Matching Person and Technology (MPT)*. In I. E. Asher (Ed.). *Occupational Therapy Assessment Tools: An annotated index* (p. 104-105). Bethesda, MD: The American Occupational Therapy Association, Inc.
- IFOPA. (2009b). *FOP fact sheet*. Retrieved from <http://ifopa.org/en/fop-fact-sheet.html>
- IFOPA. (2009d). *FOP symptoms*. Retrieved from <http://ifopa.org/en/symptoms.html>
- Kaplan, F. S., Le Merrer, M., Glaser, D., Pignolo, R. J., Goldsby, R. E., Kitterman, J. A., ... Shore, E. M. (2008). Fibrodysplasia ossificans progressiva. *Best Practice & Research Clinical Rheumatology*, *22*(1), 191-205. doi:10.1016/j.berch.2007.11.007
- Kaplan, F. S., Shore, E. M., & Pignolo, R. J. (eds.). (2011). The medical management of fibrodysplasia ossificans progressiva: current treatment considerations. *The International Clinical Consortium on Fibrodysplasia Ossificans Progressiva*. 4: 1-100.
- Kielhofner, G. (2008). *Model of human occupation* (4th ed.). Baltimore, Maryland: Lippincott Williams & Wilkins.
- Kielhofner, G., Forsyth, K., Kramer, J. M., Melton, J., & Dobson, E. (2009). The model of human occupation. In E. B. Crepeau, E. S. Cohn, & B. A. Boyt Schell (Eds.), *Willard & Spackman's occupational therapy* (11th ed., pp. 446-461). Baltimore, Maryland: Lippincott Williams & Wilkins.

- 
- Martin, L. M. (2007). Assessments of occupational performance: *Occupational Therapy Psychosocial Assessment of Learning (OTPAL)*. In I. E. Asher (Ed.). *Occupational Therapy Assessment Tools: An annotated index* (p. 488). Bethesda, MD: The American Occupational Therapy Association, Inc.
- Martin, L. M. (2007). Assessments of social participation and quality of life: *Children's Assessment of Participation and Enjoyment (CAPE)*. In I. E. Asher (Ed.). *Occupational Therapy Assessment Tools: An annotated index* (p. 204-205). Bethesda, MD: The American Occupational Therapy Association, Inc.
- Mayer, M. L., White, B. P., Ward, J. D., & Baranaby, E. M. (2002). Therapists' perceptions about making a difference in parent-child relationships in early intervention occupational therapy services. *American Journal of Occupational Therapy*, 56, 411-421.
- McWilliams, J. R. (2011). Pediatric fall risk assessment tool for home health practice. *Home Healthcare Nurse*, 29(2), 98-105.
- Parker, J., N., & Parker, P. M. (2007). *Fibrodysplasia ossificans progressiva: A bibliography and dictionary for physicians, patients, and genome researchers*. San Diego, CA: ICON Health Publications.
- Piram, M., Merrer, M. L., Bughin, V., Prost, Y.D., Fraitag, S., & Bodemer, C. (2011). Scalp nodules as a presenting sign of fibrodysplasia ossificans progressiva: A register-based study. *Journal of American Academy of Dermatology, Inc.*, 64(1), 97-101. doi:10.1016/j.jaad.2010.04.025

- 
- 
- Reynolds, S., & Lou, J. Q. (2009). The Issue Is—Occupational therapy in the age of the human genome: Occupational therapists' role in genetics research and its impact on clinical practice. *American Journal of Occupational Therapy*, 63, 511-515.
- Schultz-Krohn, W. (2007). Assessments of Occupational Performance: *Child Occupational Self Assessment*. In I. E. Asher (Ed.). *Occupational Therapy Assessment Tools: An annotated index* (p. 47). Bethesda, MD: The American Occupational Therapy Association, Inc.
- Schultz-Krohn, W. (2007). Assessments of Occupational Performance: *Model Of Human Occupation Screening Tool*. In I. E. Asher (Ed.). *Occupational Therapy Assessment Tools: An annotated index* (p. 37). Bethesda, MD: The American Occupational Therapy Association, Inc.
- Schultz-Krohn, W. (2007). Assessments of Occupational Performance: *Pediatric Volitional Questionnaire*. In I. E. Asher (Ed.). *Occupational Therapy Assessment Tools: An annotated index* (p. 53). Bethesda, MD: The American Occupational Therapy Association, Inc.
- Specht, J, King, G., Brown, E., & Foris, C. (2002). The importance of leisure in the lives of persons with congenital physical disabilities. *American Journal of Occupational Therapy*, 56, 436-445.
- Strokes, R. H., & Holsti, L. (2010). Paediatric occupational therapy: Addressing parental stress with the sense of coherence. *Canadian Journal of Occupational Therapy*, 77, 30-37. doi: 10.2182/cjot.2010.77.1.5
- Taylor, R. R. (2008). *The intentional relationship: Occupational therapy and use of self*. Philadelphia, PA: F. A. Davis Company.