

New CF Infection Control Guidelines

The first infection control guidelines for CF patients were published 10 years ago in 2003. Since then, there has been new information about the possibility of cross infection of various organisms between people with CF. Therefore, the Cystic Fibrosis Foundation has assembled a group of experts to create updated infection control guidelines. We were informed of those guidelines in March, 2013. The key points of the new guidelines are:

- At any CF Foundation sponsored indoor event, meeting or office, including gatherings such as Foundation chapter committee meetings, only one person with CF may be present.
- At Foundation-sponsored outdoor events or gatherings, people with CF need to maintain a distance of at least 6 feet from each other.
- Under no circumstances shall individuals who have ever had a confirmed positive sputum culture for *Burkholderia cepacia* (*B. cepacia*) complex attend any Foundation events, meetings or offices.

You can learn more about the new infection control policy at the CF Foundation's website:

<http://www.cff.org/aboutCFFoundation/NewsEvents/3-14-Infection-Prevention-Control-Policy-Update.cfm>

A webcast on the infection control policy is located at:

http://www.cff.org/LivingWithCF/Webcasts/ArchivedWebcasts/Germs/#Infection_Prevention_and_Control_Policy_Update

Research News

In the words of Dr. Craig Gerard, pediatric pulmonologist in Boston, "ivacaftor for CF patients is like insulin for diabetics." Ivacaftor does not cure CF, but it is the first drug to actually treat the basic defect in CF. This drug was approved by the FDA in early 2012 for patients with CF who are 6 years of age and older and who have a G551D mutation. Only approximately 4% of CF patients have a G551D mutation. Fortunately, there is exciting research occurring for patients who have two copies of the F508 mutation, the most common mutation.

In patients with the G551D mutation, the CF protein, CFTR, is present in the membrane of the cell, but the protein does not function properly. Ivacaftor works by causing the CFTR protein to function more normally and transport chloride across the cell. The issue with the F508 mutation is that the majority of the CFTR protein is broken down in the interior of the cell, and no to very little protein is present in the membrane of the cell. However, Vertex Pharmaceuticals is working on other medications that would assist the protein product of the F508 mutation to be inserted into the membrane of the cell. One such medication is called lumacaftor (VX-809). In theory, the administration of lumacaftor results in the protein from patients with the F508 mutation to be inserted in the membrane of the cell, and ivacaftor activates the protein to transport chloride across the cell.

In the fall of 2012, the results of the phase 2 study of lumacaftor + ivacaftor were reported at the North American Cystic Fibrosis Conference. Patients with CF who received the drug combination had an improvement in their FEV₁ values of 9.7%. These results have prompted Vertex Pharmaceuticals to start a large phase 3 study of this drug combination in patients with CF who have two copies of the F508 mutation, are above 12 years of age, and whose FEV₁ values are between 40% and 90% predicted. This study will occur at CF Centers throughout the country.

Vertex Pharmaceuticals is developing another drug, currently known as VX-661, that will function like lumacaftor in assisting the CFTR protein to be trafficked to the membrane of the cell. In a press release on April 18, 2013, Vertex reported the results of a study of VX-661 plus ivacaftor in CF patients with two F508 mutations. The subjects in the study who received combination therapy had a relative increase of FEV₁ of 9%. To read about the VX-661/ivacaftor study results, go to:

<http://www.cff.org/aboutCFFoundation/NewsEvents/4-18-Phase-2-Combo-Study-Kalydeco-VX-661-Results.cfm>

Save the Date

Thank you to everyone who attended the November 10, 2012 CF Family Education Day. We value the evaluations completed by the attendees and we will be using those evaluations to plan our next CF Family Education Day. For those of you who could not attend the CF Family education day, you can watch streaming videos of the presentations at: <http://videos.med.wisc.edu/events/206>

Our next Cystic Fibrosis Family Education Day will be on November 2, 2013 at the Health Sciences Learning Center (750 Highland Avenue; adjacent to the University of Wisconsin Hospital in Madison). We will post details on our CF Center website (www.uwcfcenter.org) as they become available.

University of Wisconsin Center Specific Outcomes

We and the Cystic Fibrosis Foundation agree with the concept of data transparency. This means that the averaged outcomes of patients seen in our Center should be available to patients and families. There is publicly available data on all Cystic Fibrosis Centers available at the Cystic Fibrosis Foundation website (www.cff.org). Data on that website is adjusted for attained age of patients, gender, pancreatic sufficiency, race/ethnicity, socioeconomic status, and age of diagnosis. As of the writing of this newsletter, the CFF website has not yet been updated to include the 2012 data. We expect that data to be publicly available in the near future. In the meantime, we now have our UW CF Center data for last year, and what follows in this article is the raw data reported to us for the calendar year 2011.

Pediatric Center

A very useful measure of lung function is the FEV₁ percent predicted. For patients 6-12 years of age, the University of Wisconsin CF Center median FEV₁ percent predicted is 98.7%,

which is 1.4% above the national average of 97.3%. For patients 13-17 years of age, the median FEV₁ percent predicted is 92.9% at UW compared to a national rate of 90.3%. For patients 6-17 years of age, the UW median FEV₁ percent predicted is 96.3% compared to a national average of 94.5%.

Our nutritional outcomes are expressed in terms of BMI (Body Mass Index) percentile. For pediatric patients 2-19 years of age, we strive for a BMI percentile of at least 50th percentile. The median BMI percentile for CF patients 2-19 years of age at UW was 56.8 compared to a national average of 51.4.

Adult Center

For patients 18-29 years of age, the median FEV₁ percent predicted at UW is 76.4% compared to a national average of 71.5%. For CF patients > 30 years of age, the median FEV₁ percent predicted at UW is 51.5% compared to a national average of 57.5%.

In adults over 20 years of age, one no longer uses BMI expressed as a percentile. In adult patients, BMI is expressed as the actual value with the units of kilograms per meter squared. The Cystic Fibrosis Foundation goal is that males should have a BMI of greater than or equal to 23 kilograms per meter squared and females should have a goal BMI greater than or equal to 22 kilograms per meter squared. In the UW Center, the percentage of adult patients who met those goals was 56.8% compared to the national average of 51.4%.

Trends over the last three years

The following table shows data for the past three years. The UW CF Center specific data is shown first, followed by the national average in parenthesis. This allows you to see trends that are occurring both on a local and a national level.

	2011 UW (national average)	2010 UW (national average)	2009 UW (national average)
Median FEV ₁ % predicted 6-12 years	98.7 (97.3)	102.3 (97.0)	100.9 (96.7)
Median FEV ₁ % predicted 13-17 years	92.9 (90.3)	90.8 (90.0)	92.0 (89.2)
Median FEV ₁ % predicted 6-17 years	96.3 (94.5)	96.0 (94.0)	97.8 (93.7)
Median FEV ₁ % predicted 18-29 years	76.4 (71.5)	66.3 (70.0)	60.4 (70.0)
Median FEV ₁ % predicted >30 years	51.5 (57.5)	56.3 (56.0)	50.7 (55.8)
Median BMI percentile 2-19 years	56.8 (51.4)	56.0 (49.4)	54.8 (48.9)
Adults over 20 years of age: % males with BMI >23 and females BMI >22	46.2 (44.9)	43.9 (43.3)	45.1 (42.9)



Welcome to the Cystic Fibrosis Foundation — Wisconsin Chapter, Madison Office. The Cystic Fibrosis Foundation (CFF) is the world's leader in the search for a cure for cystic fibrosis (CF), and we fund more CF research than any other organization. Nearly every CF drug available today is possible because of Foundation support. We are one of 70 chapters and branch offices and work in conjunction with the CFF — Wisconsin Chapter, Milwaukee Office. Our mission is to assure the development of the means to cure and control CF, and to improve the quality of life for those with the disease. The Cystic Fibrosis Foundation accredits 115 CF Care Centers nationwide, including the one at the American Family Children's Hospital.

We work closely with CF families, community volunteers, and corporate leaders throughout the state to host a variety of special events to raise funds for vital CF programs. In 2012, our state raised \$1,896,000 (net). We hope to increase this amount to \$1,978,000 (net) in 2013. Nearly 90% of all funds raised go directly toward our mission.

Meet The Staff



Danelle O'Neill, Executive Director

Danelle joined the Foundation in 2010. As Executive Director of the state, she oversees both the Milwaukee and Madison offices. She is implementing the inaugural Xtreme Hike in 2013 and also manages the Annual Fund, Advocacy, Major Gifts and Chapter Board of Directors. Her email is: doneill@cff.org.



Kathleen Ruby, Development Director

Kathleen joined the Foundation in 2011. She directs Great Strides Eau Claire, Great Strides La Crosse, Great Strides Stevens Point, Great Strides Wisconsin Dells and the Wine Opener. Her email is: kruby@cff.org.



Alyssa Greiber, Development Manager

Alyssa joined the Foundation in 2010. She directs Great Strides Madison, Great Strides Marshfield, Great Strides Monroe, CF Cycle For Life and the Multistack 65 Roses Golf Classic. Her email is: agreiber@cff.org.

We invite you to call, email, or visit us at our office in Madison for a personal introduction to our staff, and more information on the work that we do. We look forward to meeting you!

Cystic Fibrosis Foundation — Wisconsin Chapter, Madison Office
2990 Cahill Main, Suite 202
Madison, WI 53711
(608) 298-9902

Summer's almost here!

Helpful Tips on Staying Healthy and Remaining Active Over the Summer

After a long winter, the warm weather's finally here! Soon, school will be out for the summer, bike paths will be crowded, basketball courts packed, and swimming pools will be full of people soaking up the much-needed warm weather! As the temperature rises, people with CF should take extra care to make sure salt, energy and fluid intake is adequate before, during, and after physical activity- especially when dehydration is of concern.

Summertime Activities

Physical activity is great for all people with CF! Activities like running, biking, and swimming are great ways to break up mucus and strengthen lungs! Although people with CF can usually do everything their peers without CF can do, they are at a greater risk of dehydration and losing lots of salt while they sweat. Upset stomach and tired muscles are a result of dehydration and sweat loss, causing children and teens to be sidelined when they'd rather be out being active with their friends!

Hydration Tip: Pour a sports drink (like Gatorade) into ice cube trays. After they freeze, the Gatorade ice cubes can be added to a sports drink for an extra cold (and extra power-packed) treat to keep the body strong!

Salt Tip: Have high salt foods like pretzels, chips, and salted nuts readily accessible. Whether the snacks are kept in a bag pack, gym bag, or purse, etc., they will quickly replenish salt that's been lost while sweating.

Reminder: Salt loss and dehydration include the following signs and symptoms:

- Nausea/vomiting, muscle cramps, fatigue, headaches, salt crystals on the skin. If symptoms are experienced, stop physical activity, rest, and replenish with salty snacks and sports drinks.

Menu Planning

Summer break means that many people with CF will be home alone- but don't let being home alone interfere with getting enough to eat! Children and teens are growing a lot, and although getting enough calories to support growth is important, finding time to cook may be tough for someone who's staying home alone. Meal planning and having quick and easy items on hand is a great way to make tasty meals that provide enough energy to ride bikes, swim, go for a jog, or even just to walk around the mall! Fueling up before and after activities will give people with CF the energy they need to hang out with their friends, and feel great doing it.

Meal Planning Tip:

Remembering when to eat, coming up with meal/snack ideas, and remembering to take enzymes can be tough, but keeping a daily log can make it easy!

Enzyme Tip: Put a sticky note with a "Take enzymes" reminder stuck to the meal plan. In addition, put reminders in familiar places: a sticky note on the fridge, in the silverware drawer, on the plates and bowls in the cupboard-anywhere! Also, keeping enzymes visible and easily accessible (set out on the counter, kitchen table, etc.) can be a great visual reminder to remember to take them.

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Home alone and nothing to eat? Make this Amped-Up Mac and Cheese Dish

Ingredients

- 1 (7.25 oz) package of macaroni and cheese dinner.
- ¼ cup butter
- ¼ cup heavy cream
- 2 tablespoons powdered milk
- ½ cup shredded cheddar cheese

Directions

1. Cook the macaroni noodles in boiling water until soft. Drain the water. Return the noodles to the pot.
2. Stir in butter, cream, powdered milk, cheese, and the cheese sauce from the macaroni and cheese dinner package.
3. Reduce heat to low and mix well until the cheese has melted. You can add cooked vegetables, sliced hot dogs, or crispy bacon to this recipe.

Serving Size

3 servings: 560 calories per serving, Fat: 32 g, Sodium: 603 mg, Protein: 16 g, Calcium: 305 mg

Vitamin News!

Many of you are used to taking Source CF ABDEKs in liquid, chewable, or gel cap form. Unfortunately, the company that makes these vitamins is no longer going to be doing so. There are now two options for CF specific multivitamins: Vitamax (liquid and chewable) and Aquadeks (liquid, chewable, and gel caps). They both provide the fat soluble vitamins A, D, E, and K as well as vitamin C, many B vitamins and zinc. We hope you find these vitamins to be an acceptable substitute. We also thank you for your patience as we have been in a "vitamin shortage" with the exit of the Source CF ABDEKs from the market.

You Can Help Us Find a Cure for CF

Elsewhere in this newsletter, we discussed some of the exciting research with drugs such as ivacaftor and lumacaftor. Research studies with those drugs have occurred at other CF Centers. Although we expressed an interest in participating in clinical drug trials, our center was not selected for such studies. This lull in new studies is, fortunately, coming to an end, and the University of Wisconsin Cystic Fibrosis Center will be participating in many new studies over the next few years.

The Cystic Fibrosis Foundation has a Therapeutic Pipeline of drugs that have already been approved and others that are under investigation.

Note that there are multiple medications in development. You can help us find a cure for CF by participating in clinical trials. To highlight the importance of CF patients asking, learning and joining clinical trials, here is a message from the CEO of the CF Foundation, Dr. Robert Beall:

Dear Friends,

For more than 50 years, the Cystic Fibrosis Foundation has dedicated itself to advancing cystic fibrosis (CF) research, treatment and care. Since the CF Foundation was established in 1955 by devoted parents who wished to improve the lives of their children, we have made tremendous progress in treating this disease.

You are the key to continuing this progress. People with CF who join clinical trials are among our most important contributors to the CF research effort. You play a critical role in the development of new treatments and the search for a cure.

Clinical trials that test potential new drugs and therapies in people with CF are a major part of CF research. Drugs that are now widely used in the treatment of CF—such as Pulmozyme®, Cayston®, TOBI®, azithromycin and hypertonic saline—would not be available to all people with CF if a small group of people with CF had not first taken part in clinical trials.

We are now testing more potential drugs in clinical trials than ever before, including new therapies that target the basic defect that causes CF. To help these drugs move swiftly from the research and testing stages into the hands of patients who

need them, we need more people with CF to take part in clinical trials. Without volunteers—without people like you—research and progress are not possible.

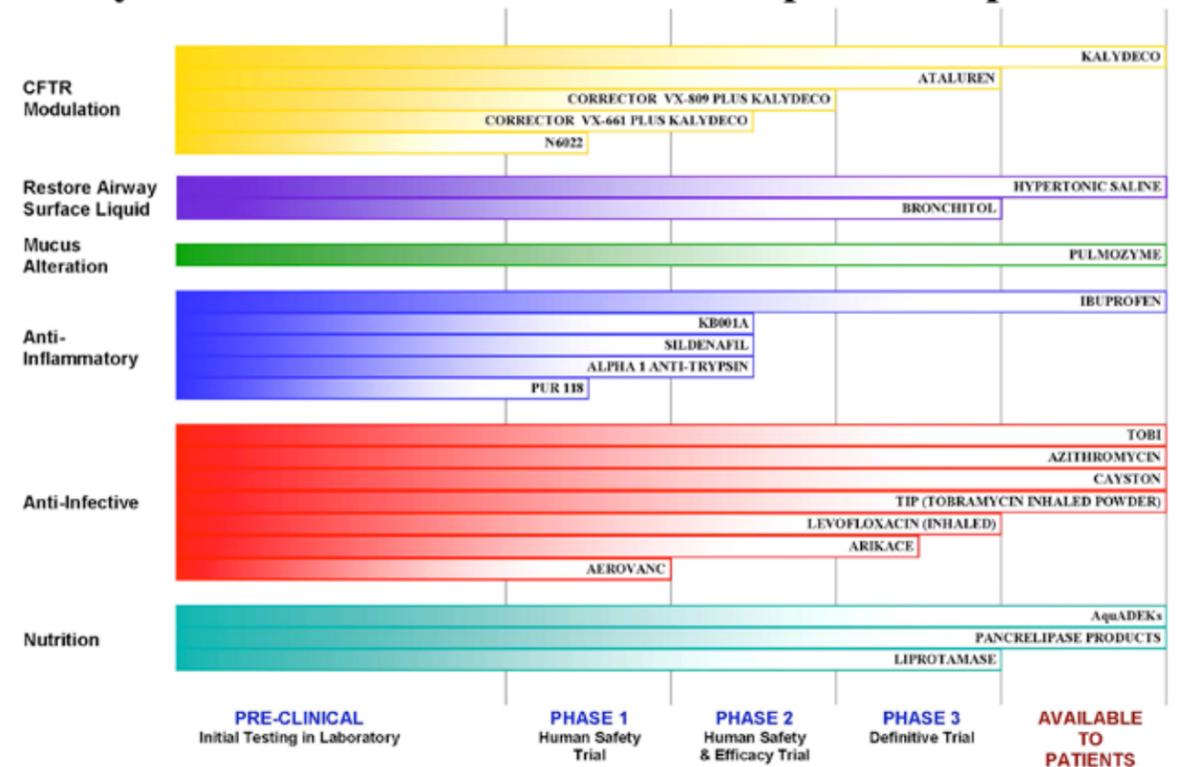
We invite all people with CF and their families to learn more about clinical research, ask their doctors and CF care center teams for information about clinical trials and join in the effort to develop new treatments by volunteering for clinical trials.

By participating in clinical trials, you help our community continue to move forward so that everyone with CF can live healthier and longer lives.



Robert J. Beall, Ph.D.
President and Chief Executive Officer
Cystic Fibrosis Foundation

Cystic Fibrosis Foundation Therapeutics Pipeline



News from the Cystic Fibrosis Foundation – Madison Chapter

On April 19, 2013 Vertex Pharmaceuticals announced promising results from a Phase 2 clinical trial of Kalydeco in combination with VX-661, another potential CF therapy. The results showed a significant improvement in lung function in people with two copies of the most common CF mutation, Delta F508. Visit the www.cff.org website to learn more about this wonderful news!

The Madison Chapter of the CF Foundation strives to keep CF families and friends up to date on national CF news, chapter events and advocacy information. Our monthly newsletter is a great way to stay in touch and learn more about how to get involved in helping us find a cure for CF. Please sign up for our monthly newsletter by visiting our website at <http://www.cff.org/Chapters/madison/> and clicking on the newsletter sign up link.

WISCONSIN CONTACT INFO

Madison (608) 298-9902

www.cff.org/Chapters/madison

Milwaukee – (262) 798-2060

We hope you enjoy the “Meet the Staff” information in this newsletter. Our Madison staff cover much of Wisconsin including LaCrosse, EauClaire, Marshfield, Monroe, Stevens Point, Wisconsin Dells and of course, Madison. That is a lot of ground for three people to cover! So we are asking you to help us by getting involved as a volunteer with one of our many Chapter events. We cannot do this without you! We need help planning and recruiting participation in the following events: Cycle for Life, Xtreme Hike, The Wine Opener. Getting involved helps connect you with other CF families and new friends. Our events are a great way to learn new skills, exercise your creativity and make a difference! Please call or email us today to find out how you can get involved.

NEW EVENTS FOR 2013

We have two new events coming up and hope you **SAVE THE DATES** on your calendar.

XTREME HIKE – Saturday, October 5, 2013

Ice Age National Scenic Trail, Southern Kettle Moraine unit
Staff contacts: Danelle O’Neill (doneill@cff.org) or Kathleen Ruby (kruby@cff.org)

The Wisconsin Chapter of the Cystic Fibrosis Foundation is proud to present the 1st Annual XTREME HIKE on Saturday, October 5, 2013. In just one day, a dedicated team of Xtreme Hikers will trek 30 miles on the Ice Age Trail to raise funds and awareness for cystic fibrosis (CF), this country’s most fatal genetic disease among children and young adults. Wisconsin is the perfect place to host this event. The natural beauty of our State is unparalleled, especially in fall. We are committing ourselves to host this event each year on a different section of the Ice Age Trail until a cure is found. This will be a celebration of the beauty and bounty of Wisconsin and we will be showcasing Wisconsin food products on and off the trail!

Madison Wine Opener

Friday, November 1, 2013
Sheraton, Madison

Staff contact: Kathleen Ruby (kruby@cff.org)

Join us for this casual evening of spirits and food tasting event benefiting CFF. Madison vendors participate in this “after work happy hour” event. Live music sets a fun atmosphere for people to mingle as they walk around the venue, sampling the food and drinks, auctions, raffles and more!

GREAT STRIDES TIME IS HERE

The CF Foundation’s largest fundraising event takes place nationwide throughout May. The pledge-based walks feature scenic routes, snacks, entertainment and prizes. Teams include families, friends, schools, companies and more. Great Strides walk on Saturday, April 27, 2013
Pierce Park (Pavilion), Appleton
Wildwood Park, Marshfield

Great Strides walk on Saturday, May 4, 2013
Eagles Club, Fond du Lac
Wisconsin Dells Ducks, Wisconsin Dells

Great Strides walk on Saturday, May 11, 2013
St. Luke United Methodist Church, Sheboygan
Great Strides walks on Saturday, May 18, 2013
Carson Park, Eau Claire
Green Isle Park Pavilion, Green Bay
Myrick Park, La Crosse
Twining Park, Monroe

Great Strides walks on Sunday, May 19, 2013
Elver Park, Madison
Milwaukee Lakefront at O’Donnell Park Milwaukee
Piffner Park, Stevens Point

Cycle for Life

June 2, 2013

Capital Brewery

NEW: Join the Madison CF Cycle for Life ride and receive a complimentary 1 year membership to the Capital Brewery Bike Club! Membership includes: bi weekly organized rides starting in May, Capital Brewery hosted events, subscription to the Capital Brewery Cycling newsletter, camaraderie and so much more!

July 14 & 15, 2013 Multistack 65 Roses Golf Classic

La Crosse Country Club, Onalaska

Staff contact: Alyssa Greiber (agreiber@cff.org)

Hosted by Multistack, the 7th annual event is held at a private country club and includes 18 holes of golf with on-course contests. The evening program includes cocktails, dinner, live and silent auctions, and featured speaker.

July 22, 2013 Four-Ball Golf Challenge

Hidden Glen Golf Club, Cedarburg

Staff contact: Lola Budzinski (lbudzinski@cff.org)

Four-ball is a twist on the average golf outing offering a real tournament to challenge both scratch and avid golfers! In addition to 27 holes on this beautiful course, the day includes breakfast, lunch, cocktails, on-course contests, raffles... and a post-golf reception featuring a wonderful dinner, evening cocktails, exciting auctions and awards presentation! Along with terrific prizes and friendly volunteers, this event is truly great golf for a great cause.

September 23, 2013 Milwaukee Bucks Golf Tournament

Westmoor Country Club, Brookfield

Staff contact: Lola Budzinski (lbudzinski@cff.org)

Join us for one of Milwaukee’s premiere golf events filled with Milwaukee Bucks players, coaches and other local sports and media celebrities! The day, which includes an 18-hole scramble golf format, begins with a noon shotgun start. Post-golf, evening festivities include a dinner reception followed by a short question and answer with the Coach and GM. As always, we’ll have incredible sports memorabilia, trips and other unique items in our auctions.

October 19, 2013 Breath of Life Gala

River Club of Mequon

Staff contact: Kelly Salentine (ksalentine@cff.org)

The Breath of Life Gala is the Milwaukee area’s premier event. This exciting black-tie-optional gala is unlike any other! Throughout the evening, guests will enjoy a cocktail reception, sit-down dinner, participate in a silent auction and enjoy other entertainment and surprises.

November 1, 2013 Madison Wine Opener

Sheraton, Madison

Staff contact: Kathleen Ruby (kruby@cff.org)

Join us for this casual evening of spirits and food tasting event benefiting CFF. Madison vendors participate in this “after work happy hour” event. Live music sets a fun atmosphere for people to mingle as they walk around the venue, sampling the food and drinks, auctions, raffles and more!

November 14, 2013 Climbing for a Cure

US Bank Building, Milwaukee

Staff contact: Julie Brzostowicz (jbrzostowicz@cff.org)

Climbing for a Cure is a pledge-based event in which approximately 500 people meet at the US Bank Building, the tallest building in Milwaukee, to face the footrace of a lifetime: 47 floors, 94 flights, 1,034 steps – straight up! Once participants reach the top, they have the opportunity to enjoy amazing panoramic views of downtown Milwaukee before heading down to the post-climb celebration complete with food, drinks, entertainment and awards! Participants include, but are not limited to, corporate teams, local fire and police departments and fitness fanatics.