The HCU Herald

In This Issue

Recipes from the Kitchen w/Amber Gibson

Did you know that almost every day has a national celebration of something? February alone has 106 national days of celebration (https://nationaldaycalendar.com/february/) and 40 of them have to do with food or drink? So, while we are all familiar with Valentine's day, President's day, and Rare Disease day – we bring to you some lesser known national days of celebration and inspired recipes to go with them.

February 4 is National Soup Day
February 10 is National Cream Cheese Brownie Day
February 20 is National Cherry Pie Day

French Onion Soup

Ingredients
622 g Slivered Onions
6 TBSP Butter, regular or unsalted
3 c Vegetable Broth
1/2 c Wine, Red
1 1/2 c Water
1/4 c Coconut Aminos
7/8 fl.oz. Recipe Secrets Onion Mushroom Soup & Dip Mix, dry, 1 packet
1 tsp Thyme, fresh, about 2 sprigs
1 tsp Bay Leaf, one leaf
1 TBSP Chopped Fresh Rosemary

Directions

1. In a large skillet melt the butter over medium heat. Once the butter begins to bubble a little, add the onions. Continue to sauté until the onions are nice and caramelized, about 20 minutes or so. Be sure to stir frequently to avoid burning any of the onions. If the onions seem to be dry, add a little extra butter to prevent sticking.
2. While the onions are cooking, combine all other ingredients into a slow cooker. Once the onions are done cooking, add them to the slow cooker with the broth mix then cover with the lid. Turn on the slow cooker to high and cook for 4 hours. Season with salt and pepper, if desired.
3. To serve, you can top with low protein croutons, then add a slice of nondairy cheese on top and broil to melt cheese. Enjoy!

Notes:
May serve topped with cheese and/or serve with crackers – not included in nutritional value.

Brownie Cookies

Ingredients

225 g Taste Connections Multibaking Mix
40 g Chocolate Pudding, dry mix only
1 tsp Baking Soda
1 TBSP Cocoa Powder
1/2 tsp Salt, Table
8 TBSP Butter, softened
1/2 c Sugar, White Granulated
1/2 c Sugar, Brown, packed
1 tsp Vanilla Extract
25 g Lightly Mixed Large Egg
2 TBSP Water

Directions

1. Preheat oven to 375 degrees. Line a cookie sheet with parchment paper and set aside.
2. In a medium bowl combine the multibaking mix, chocolate pudding mix, baking soda, cocoa powder, and salt. Lightly whisk to combine. Set aside.
3. In the bowl of a stand mixer add the softened butter and the sugars and mix together until light and fluffy. Add the vanilla and mixed egg to the sugar mixture just until combined. Add half of the dry ingredients to the wet ingredients mixing just until combined. Add the other half of the dry ingredients and mix just until combined. Add one or two tablespoons of water and mix until combined.
4. Use a cookie scoop and place dough balls onto prepared cookie sheet, two inches apart. I got 12 dough balls on my cookie sheet. Bake in oven for 10 to 12 minutes until edges are lightly browned. Do not over bake. Remove from oven and allow to cool on cookie sheet for 5 minutes before removing to a cooling rack

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Servings: 25
Serving size: 1 cookie
Protein per serving: 0.3 g
Calories per serving: 108

Cherry Goodness Cookies

Ingredients

290 g Wheat Starch

1 tsp Xanthan Gum

1/2 tsp Salt, Table

50 g Vanilla Pudding, dry mix only

12 TBSP Butter, regular or unsalted, softened

1/2 c Sugar, Brown, packed

1/2 c Sugar, White Granulated

25 g Mixed Large Egg, mix the egg first, then measure

1/2 tsp Imitation Cherry Extract, up to 1 tsp if desired

2 TBSP Maraschino Cherry Juice

80 g Finely Chopped Maraschino Cherries

50 g Semi-Sweet Chocolate Chips

*1 tsp Pink or Red Food Coloring*Directions

1. Combine all dry ingredients in a medium bowl and lightly whisk to combine. Set aside.
2. In the bowl of a stand mixer, add the butter and sugars. Cream together until light and fluffy. Scrape bowl then add the egg, cherry flavoring, and maraschino cherry juice. Mix just until combined. Add the dry ingredients, in two batches, and mix just until combined. Add the food coloring now and gently mix until desired color is reached. Fold in the cherries and chocolate chips. If dough seems a little thick, add one tablespoon of water and gently mix together. Refrigerate dough for about 20 minutes.
3. While dough is chilling, preheat oven to 375 degrees and line a cookie sheet with parchment. Take cookie dough out and use a cookie scoop to place dough on baking sheet. Be sure to space each scoop about 2 inches apart, as these cookies may spread a little. Bake for 12 to 15 minutes until cookies are set and edges are browned a little. Allow to cool for at least five minutes before moving to a cooling rack, as these will be very soft.

Servings: 26
Serving size: 1 cookie
Protein per serving: 0.3 g
Calories per serving: 142

About Rare Disease Day
Rare Disease Day® takes place on the last day of February each year. The goal of Rare Disease Day is to raise awareness amongst the general public and decision makers about rare diseases and their impact on patients' lives. It's never too early to get involved! The National Organization for Rare Disorders (NORD) is the official Sponsor of Rare Disease Day in the United States. To learn what's happening around the globe, visit the Rare Disease Day website at rarediseaseday.org.

What is a Rare Disease?
In the U.S., any disease affecting fewer than 200,000 people is considered rare. This definition comes from the Orphan Drug Act of 1983 and is slightly different from the definition used in Europe. There are more than 7,000 rare diseases affecting 25-30 million Americans. In other words, one in ten Americans are suffering from a rare disease and more than half of them are children. Besides dealing with their specific medical problems, people with rare diseases struggle to receive a proper diagnosis, find information and get treatment. The rarity of their conditions makes medical research more difficult.

Show Your Stripes™
The zebra is the official symbol of rare diseases in the United States and is noted for its black and white stripes, which are central to its uniqueness. Everyone has his/her own stripes, those characteristics that make each individual distinct. While each of the more than 7,000 rare diseases are unique, there are many commonalities that unite patients, families, caregivers and supporters. In the spirit of raising the profile of the rare disease community at large and celebrating Rare Disease Day, this year NORD is promoting a variety of ways in which individuals, organizations and groups can "show their stripes."

Raise Social Awareness
HCU Network America will be sharing a fact each day about Homocystinuria on our social media accounts. We encourage you to share our post. In addition to sharing our post, we encourage you to check out the NORD Social Media Toolkit

What does rare mean to you ?

During February, leading up to Rare Disease Day, we challenge you to write an Acrostic Poem about what Rare means to you.

An acrostic poem is a poem where certain letters in each line spell out a word or phrase. Typically, the first letters of each line are used to spell the message, but they can appear anywhere.

Remember to use the hashtags #ShowYourStripes , #RareDiseaseDay #GoBlueforHCU

Here is our example:

What does rare mean to you?
Rarely recognized by its name
Always needs more Awareness
Requires low protein diet
Explanations guaranteed

Rare Disease Day - Casual for a Cause

Itching to forgo the traditional slacks and skirts for a more relaxed look at the office? Take the lead in your organization or company to sponsor a Casual Cause: Jeans Day to raise funding for the resources and tools HCU Network America provides to the Homocystinuria community! Encourage employees to dress down for a day or even an entire week by requesting donations in exchange for a day in their casual best.

Not sure where to start? Not a problem! We've created several "Team Captain" materials to help you on your way toward sponsoring a successful fundraiser. Take a look at our customizable flyers, stock emails to inspire participation, tracking sheets and more! View resources at: https://hcunetworkamerica.org/casual-for-a-cause/

For more information, please contact Danae' Bartke at 630-360-2087
or dbartke@hcunetworkamerica.org

Texas HCU Family Picnic

Registration now open: https://hcunetworkamerica.org/texas-family-picnic/

Stay tuned! Want to make sure you are notified right away when we have the details and registration opens? Email us – dbartke@hcunetworkamerica.org

Aeglea Presents at JP Morgan 38th Annual Healthcare Conference

January 15, 2020 Aeglea BioTherapeutics' CEO, Anthony Quinn, presented at the 38th JP Morgan Annual Healthcare conference in San Francisco, California. Aeglea is a biotechnology company developing next-generation human enzyme therapies for rare diseases.

At the conference they talked about who Aeglea is, what makes them unique and what their clinical pipeline looks like. They currently have 3 therapies in development with their lead therapy being Pegzilarginase, which is in phase 3 clinical trial for Arginase 1 Deficiency. They presented data and video, which shows this therapy is having impeccable outcomes.

They then spoke about their other programs, ACN00177 for Cystathionine Beta-Synthase (CBS) Homocystinuria (HCU) and AEB5100 for Cystinuria. Regarding CBS HCU, Dr. Quinn spoke about the incidence rate and symptoms for undiagnosed and/or untreated HCU, and the inadequacies in current disease management. He then presented data from their mouse model, showing they have 100% survival and improvements in disease related abnormalities. Dr. Quinn then outlined the clinical trial process. They have filed a Clinical Trial Application in the UK and Investigational New Drug Application in the US and expect Phase 1/2 clinical trial to be initiated in the 2nd quarter of 2020, with initial clinical data expected in the first quarter of 2021. HCU Network America is thrilled to see another potential therapy moving forward in development for HCU!

To view the recording, please visit: https://jpmorgan.metameetings.net/events/hc20/sessions/29703-aeglea-biotherapeutics

Finding your Local "Village"

Rare Disease Day – "Alone we are Rare, together we are Strong" – NORD

HCU Network America is just one of many organizations across the country that help provide community to those with Homocystinuria. Here at HCU Network America, we firmly believe patients and their caregivers are more successful when they have a community to support them, not just online, but locally (or regionally) as well. Across the country many PKU organizations include what they call Allied Disorders. Allied disorders include other amino acid based disorders that require a low protein diet, such as classical Homocystinuria.

Arizona Network for PKU and Allied Disorders (ANPAD)
ANPAD's mission is to help individuals with PKU and Allied Disorders adhere to their treatment, inform families of the latest research and treatment advances, raise public awareness of PKU and Allied Disorders and create a support network. This is accomplished through camps, cooking workshops, educational seminars, online and print publications and newsletters, and recreational activities which bring families together to learn, share ideas, and encourage one another. Website: https://anpadnews.org/
Twitter: @azpku | Facebook: anpadnews | Youtube: livingwithpku |Instagram: azpku

California Coalition for PKU and Allied Disorders (CCPKUAD)
CCPKUAD is a 501(3)c non-profit organization. We are a volunteer group of individuals committed to provide support, information, education and advocacy to individuals with PKU and other inborn errors of metabolism. https://ccpkuad.org | Facebook: Ccpkuad | Twitter: @CCPKUAD |Instagram: ccpkuad_

Georgia PKU Connect
https://georgiapku.org | Facebook: GeorgiaPKU | Instagram: georgiapku | Twitter: @GeorgiaPKU_USA

PKU Organization of Illinois and Allied Disorders
We have three key parts to our mission for the PKU Organization of Illinois. First, and one of the most important, is to support the person, and their families affected by PKU and/or Allied Disorders. Second, is the organization will spread information and awareness of PKU and Allied Disorders to the PKU/Allied Disorder community, as well as, the outside community. Lastly, constantly raising funds to aid in the continued research
of medical updates and changes of PKU and Allied Disorders. As part of our fund raising, we will stay connected with local and national lawmakers to be informed and inform others of the medical changes and necessary events related to PKU and Allied Disorders. https://pkuil.org | Facebook: PKUIllinois |Twitter: PKUIllinois | Instagram: pkuillinois | YouTube: https://www.youtube.com/channel/UCtb-na6h59liGg3LKNHkeoiQ/featured

Intermountain PKU and Allied Disorders (IPAD)
The Intermountain PKU and Allied Disorder's mission is to provide support and services to individuals and families with PKU and allied disorders, to encourage research, and to increase public awareness of these conditions. http://ipadutah.org/ | Facebook: IntermountainPKU Instagram: Intermountain_pku | Twitter: @IntermountainP

Michigan PKU & Associated Disorders
The mission of the Michigan PKU Organization is to provide support and education to individuals and families affected by PKU and similar metabolic disorders, raise community awareness, support PKU research, and promote the overall health and well-being of Michiganders living with PKU and similar metabolic disorders. https://michiganpku.org
Facebook: MichiganPKU | Instagram: pkumichigan

The Mid-Atlantic Connection for PKU and Allied Disorders
The Mid-Atlantic Connection for PKU and Allied Disorders (MACPAD) is a non-profit 501(c)(3) organization, dedicated to improving the health and wellbeing of individuals and families affected by Phenylketonuria (PKU) and related metabolic disorders. The mission of MACPAD is to enrich the lives of individuals and families of individuals with inherited metabolic disorders by disseminating information, providing supportive activities and encouraging the exchange of ideas. http://www.macpad.org/home | Facebook: joinMACPAD Instagram: macpad_upstate_ny

New England Connection for PKU and Allied Disorders (NECPAD)
The New England Connection for PKU and Allied Disorders, Inc. (NECPAD) supports families with PKU, HCU and Allied Disorders in Connecticut, Rhode Island, Massachusetts, Vermont, New Hampshire, and Maine. NECPAD supports individuals and families with financial assistance, up-to-date information regarding new therapies and medical treatments, scholarships, networking, and social events. www.necpad.org| Facebook: necpad |Twitter: @NECPADOrg | Instagram: necpadorg

PKU Northwest
Providing support, networking, and community engagement to Northwest families and individuals with PKU and Allied Disorders. https://pkunw.org/ | Facebook: pkunorthwest | Instagram: pkunw

Tennessee PKU Foundation
The mission of the Tennessee PKU Foundation is to provide support and education to individuals and families affected by PKU and similar metabolic disorders, raise community awareness, support PKU research, and promote the overall health and well-being of Tennesseans living with PKU and similar metabolic disorders. http://tennesseepku.org/ | Facebook: TNPKUFoundation | Instagram: tnpkufoundation

Louisiana Metabolic DisordersCoalition
The mission of the LMDC is to support, educate, and advocate for patients and families that are affected by metabolic disorders. Facebook: https://www.facebook.com/louisianamdc/

Organic Academia Organization
We are a volunteer non-profit organization whose mission is to empower families and health care professionals with knowledge in organic acidemia metabolic disorders. We support early intervention through expanded newborn screening, solicit contributions and distribute funding that supports research toward improved treatment and eventual cures in the areas of Organic Acid disorders. https://www.oaanews.org/ | Facebook: OrganicAcidemiaAssociation | Twitter: organicacidemia

HCU Heroes

Derek

From Pennsylvania

Derek was diagnosed with HCU during the newborn blood screening, so we were lucky enough to have been able to start treatment from birth. As for most parents, the initial phone call we received was shocking and filled with many unanswered questions. We received the call from our local lab who said there was an issue with Derek's labs and we needed to set up an appointment at our local Children's Hospital. The following week we were seen at Children's Hospital in Philadelphia (CHOP) by the genetics team. Many of our questions were still not answered at this appointment, but we were told to start him on a special formula and that he was not able to break down protein. We did some research on the internet which was a big mistake as most people know this can cause a lot of fear (my best advice - do not google HCU). We do have a good team at CHOP and they were able to provide us with some reassuring answers and have been very supportive ever since.

Derek is now 16 and will turn 17 in February. He does not talk about the disorder and he keeps it very hush, hush. We struggle with getting him to take his medicine and follow his diet. To date I am very thankful he has not had any medical issues, but I worry daily about the side effects he may experience because he does not take HCU serious. His levels are never at a normal range and for the most part are extremely high. At his age, he needs to be accountable and it is very hard to make him understand that. He is very involved in sports and is very athletic. As parents, we are extremely thankful he leads a normal life, and outside of the diet and medicine he is very lucky. Trying to get him to understand the long term effects has been our biggest challenge. I have recently become part of the HCU Network America group on Facebook and the support from there has been a great help to me. Before that it was just me researching and not having any support or others to talk about his issues. We met some of the members over the summer in Atlanta and that was an incredible experience for me. I hope one day as he matures, he will find support and help in the group as well. Explaining the disorder is also challenging because of how rare it is. Derek is also in the new OT-58 study and is part of the second group. We have not had much feedback and have no idea if it is helping or not. But the nurse comes weekly and we go to CHOP every 6 weeks. We pray this may be the answer patients have been looking for but only time will tell. He still has about another year in the study. Our story is pretty simple but nonetheless I wanted to get his story out there. All of the support is appreciated even if he does not want to participate; it helps my peace of mind.

Medical Nutritional Equality Act

Kylie Barber, MS, MJ

"Our family is unique, in that I have [an inherited metabolic disorder] as well as our four adopted sons. With no guarantee of insurance coverage, I fear that my children will one day be forced into a situation similar to that they were born into: their birth families lived in a country where medical care was not affordable and they had to abandon their children, hoping they could be raised under better conditions. One son was not treated for his medical condition for the first 13+ years of his life in his birth country. The result: impaired cognitive function (much of it is irreversible)."

Each year, thousands of children and adults in the United States are diagnosed with certain digestive or inherited metabolic disorders that prevent their bodies from digesting or metabolizing the food they eat. For them, medically necessary food is the standard of care treatment. Due to the cost of treatment and lack of coherent insurance coverage, many individuals and families struggle to gain access to their life-sustaining treatment. Unfortunately, the story above is not unlike many other stories of families who struggle with an inborn error of metabolism (IEM) diagnosis. The typical worries that are concomitant with raising a family are prodigiously exacerbated by this diagnosis and many families are forced to choose between their child's health and the overall well-being of their family.

The Medical Nutrition Equity Act (MNEA), sponsored by Congressman Jim McGovern (D-MA) and Congresswoman Jamie Herrera-Beutler (R-WA) in the U.S. House of Representatives (and to be introduced by Senator Bob Casey (D-PA) in the Senate later this year) would be lifechanging and lifesaving for IEM patients. This legislation provides for the coverage of medical formula and low-protein modified food products, as well as, vitamins and individual amino acids for digestive and inherited metabolic disorders under Federal health programs and private insurance so long as it is pursuant to the prescription or order of a physician.

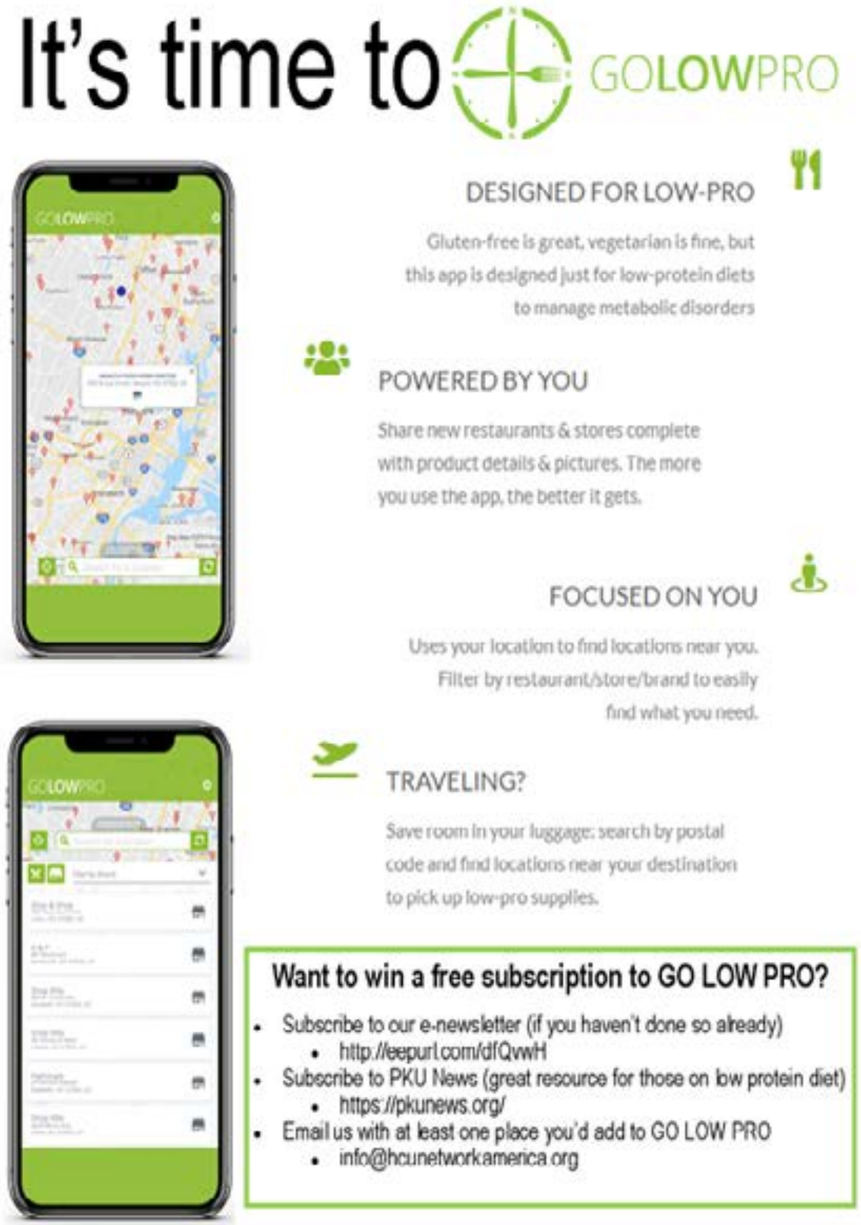
Since its reintroduction into the U.S. House of Representatives in early May 2019, the Medical Nutrition Equity Act (H.R. 2501) has shown tremendous movement. The legislation currently has a record-breaking 52 cosponsors, quadruple the number this time last Congress. The success of this bill is majorly attributed to the grassroots advocacy efforts of the patient community. While there is not necessarily a desired "magic number" of cosponsors, the more Congressional support this legislation has, the more quickly it will progress through the legislative process to become law.

During Rare Disease Week 2020 and Rare Across America 2020, many patients and advocates will meet in person with their Members of Congress and/or staffers to advocate and lobby for the Medical Nutrition Equity Act. These initiatives will be organized by the Rare Disease Legislative Advocates (RDLA), a program of the EveryLife Foundation for Rare Diseases. Reporting on her Rare Across America 2019 meeting with Congressman McGovern, MNEA advocate Anne said, "Thank you for putting these meetings together. Being involved with Rare Disease Week in DC and these summer events has been transformative for me. Suffering from rare diseases can feel disempowering and getting involved with RDLA this year has given me hope and agency."

The current priority of the Medical Nutrition Equity Act is to garner as much legislative support as possible by way of building cosponsors on H.R. 2501. The Senate version of the bill is in need of a Republican lead cosponsor before it can be reintroduced. While there are currently four potential Republican leads, including Senators Bill Cassidy (LA), Joni Ernst (IA), James Lankford (OK), and Steve Daines (MT), the timeline for sponsorship is unknown.

For health care professionals interested in becoming involved in advocating for the Medical Nutrition Equity Act, now is the time, more than ever, to act! With record-breaking numbers and vigorous grassroots engagement, exciting changes are happening. Act now by sending an email to your Member of Congress urging them to sign on as cosponsor of H.R. 2501 by clicking here! Patients are also encouraged to share their story on the Patients and Providers for Medical Nutrition Equity Coalition website.

For questions or more information about advocacy opportunities, contact Kylie Barber at KBarber@Everylifefoundation.org.



Traveling with HCU: Bharat Ramanujam

VISIT TO DALLAS – DEC 19-21

I went to Dallas and visited the President George H. Bush Presidential museums and two other Aviation Museums. We stayed at Homewood Suites in Plano Texas, which provides cooking facilities.

Breakfast at the Hotel: Hash browns with Salsa (both green and red). More of Salsa and less of Potato (about 100 gms). Also had 30 gms of Fruit Loops cereal with Unsweetened Almond Milk.

Packed Lunch: Potato, salsa and low-pro bread

Dinner: Visited an Indian vegetarian restaurant, Rajula, for dinner. This restaurant provides a variety of vegetable curries, low in protein and carbs. Had tindora (mini cucumber) and cabbage curries and 2 pieces Green Chili Tempura (has small amount of flour), with my low-protein tortillas.

VISIT TO ORLANDO – DEC 21-25

In Orlando we visited the Kennedy Space Center, Fantasy of Flight Air Museum. While in Orlando, we met the astronaut Sherwood Springs. Stayed at Homewood Suites in Lake Mary near Orlando, which provides cooking facilities.

Breakfast at the Hotel: Hash browns with Salsa (both green and red). More of Salsa and less of Potato (about 100 gms). Also had 30 gms of Fruit Loops cereal with Unsweetened Almond Milk.

Packed Lunch: Potato and low-protein sandwich for lunch.

Dinner: Visited Indian vegetarian restaurant, Woodlands, for dinner. This restaurant provides a variety of vegetable curries low in protein and carbs. I had eggplant and mixed vegetable curries and also a small amount of different starters, samosa (pastries with potatoes), green chili tempura, with my low-protein tortillas

Visited a Pizza place: Pizza@ in Titusville (near Kennedy Space Center). They serve Gluten Free Pizza.

VISIT TO HOUSTON – DEC 28-JAN 2

On the way to Houston, we stopped in Austin and visited the Lynden B. Johnson Presidential Museum. While in Houston, we went to the Johnson Space Center. The highlight was the visit to the Historic Control Center which was used for many Gemini, Apollo and Space Shuttle missions, including the famous Apollo 11.
Stayed at Homewood Suites in Clearlake, very close to Space Center. Bought Almond Milk and watermelon (low carb and protein), before we checked in the hotel. This hotel provides some kind of potato for me whenever we stay.
Breakfast at the Hotel: Hash browns with salsa. Also had 30 gms of Fruit loops cereal with Unsweetened Almond Milk.
Packed Lunch: Potato, salsa and low-protein bread.
Dinner: We visited a vegetarian Indian restaurant called Bombay, which has an all day buffet. I ate a lot of pickled vegetables for dinner. This restaurant also provides a variety of vegetable curries which I can eat with low-protein tortillas. I had cauliflower curry and ordered eggplant curry since it wasn't on the buffet to make it a lower protein dinner.

My health on the road:

I had 15 fractures total, 12 prior to 2011 when my Vitamin D levels were low (less than 15). I had a shoulder fracture 2 years ago when I fell hard on my right hand and it took over 10 months for it to completely heal.

In the summer 2019, while in Austria, I fell on the street and suffered a left knee hair line fracture. After my fall I was in the Salzburg hospital for 6 hours until they diagnosed the fracture and gave me a brace. Instead of enjoying my trip in Austria and Budapest, I was in a wheelchair for 3 weeks with a brace while touring around. The doctor at the hospital told me I could still walk but I suffered and had to wait until the end for me to walk without holding on to something. I was not able to go to the Salzburg Cave or Palace, and climbing the stairs in places like Mozart houses in Salzburg, Austria and other places in Salzburg and Vienna was torture.

Visiting Austria and Budapest in September was my worst trip. I was not able to walk or climb properly. I was permanently in a wheelchair for 4 weeks. It was miserable physically and mentally because of my left knee injury. In Europe I ate a lot of veggies, gluten-free pasta and bread, and vegan non-dairy cheese. It was easy to manage everywhere we went; even the pierogis in Poland with cabbage and mushrooms, without cheese, was fantastic. Airport lounges also had lots of veggie options for me.

Then within 6 weeks, on November 4th while in Boston, my second injury to my right foot, ankle and leg happened when I fell down the stairs. I had to undergo surgery 3 weeks later since there was so much swelling. Since then, I have not been able to volunteer at the California Science Center in their Endeavour Pavilion. It has been frustrating for me because I have worked there for 5 years and have donated 5,300 hours of my time.

Since November 4th, 2019 I have been going through the steps below:
1. Temporary splint
2. Several x-rays and MRIs
3. Surgery (they put screws in the left and right side of my ankle and then on the top of my foot and back fibula leg bone).
4. Temporary splint for two weeks, while healing took place.
5. Stitches removed
6. Wheelchair bound for 4 more weeks
7. Claustrophobic, non-weight baring boot for 4 weeks.
8. Currently in claustrophobic boot.

I tell every HCU patient to monitor the following levels:
- Vitamin D
- Bone Density Test
- Calcium

My Vitamin D levels are now up to 77, and I'm healing well. Hopefully keeping my Vitamin D levels high enough through taking enough natural calcium via almond milk and green vegetables and supplements from HCU Express will help me.

Ask Methia

Dear Methia,

I'm sure you hear this all the time, but this low protein diet is ridiculously hard. I'll find myself allowing one "cheat day," usually when I'm out with friends, which leads to longer periods of non-compliance. I have now been off diet for 8 months and during that time gained 25 pounds. I don't really understand how that happened, since I am still really active, and not taking my formula has reduced my intake by about 400 calories. What gives?

Sincerely,

Pizza with extra (NON-vegan) cheese

Dear Pizza,

We've all been there. You've just met someone new – a "friend-of-a-friend," a colleague, or even a first date. You tell them that you can only eat 15 grams of protein per day and their jaw nearly hits the floor. Questions follow, including, "Well, if you just ate one piece of chicken, it would be OK, right?" WRONG!

I digress, though. Let's talk about why you might have gained this weight.

• The formula that you self-discontinued? It's what helps to keep you full. Repeat after me: "Formula Is Medicine." Your medical food is formulated to give you not only the synthetic, methionine-free protein that you need, but to give you vitamins, minerals, and calories that become deficient in an overall restricted diet. Drinking your formula before a meal is the most nutritious thing your body can have, and it provides an element of satiety (that warm feeling you experience when you're full, but not too full).

• Less formula = more food (typically). It's obvious that you have WAY more options at mealtimes when not following your diet. It also makes sense that the "unrestricted" mindset can result in over-eating what your body might really need. Those fries that you usually weigh out at the restaurant are now being plucked from the basket in uncontrolled portion sizes. A caloric surplus beyond your needs will ALWAYS result in weight gain.

Slowly returning to diet – perhaps adding formula to one meal to start, and gradually eliminating higher protein foods – is the best way to get back on track. Remember, fad diets aren't good for anyone, but ESPECIALLY not for metabolic patients – that Keto/Intermittent Fasting trend that your friend is trying is downright dangerous for you and your metabolic control. More attention to portion sizes, meeting (not exceeding!) your protein goals, and adding back that formula will have you feeling better in no time.

Sincerely,
Methia

HCU Awareness T-Shirts

HCU Awareness T-Shirts are back!
Get your official 2020 HCU Awareness T-Shirts. All proceeds benefit HCU Network America!

Buy me now:
bonfire.com/hope-connects-us-letter-tiles
Pick me, Pick me!
https://www.bonfire.com/rare-disease-day-hcu/