

The PKU

PRESS

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simplified diet

WHAT IS IT
AND HOW TO
USE IT

lets chat...

TIPS FOR
MANAGING PKU
AT DAYCARE

*special event
feature!*

LIFTING THE LIMITS
FOR PKU
CHICAGO
GALA

**VARY
YOUR
VEGGIES**

OUR MONTHLY VEGGIE
SPOTLIGHT: POTATOES

note from the editor

Hello! Welcome to the reinstated newsletter, The PKU Press, published by the PKU Organization of Illinois! In this issue I share the Lurie Children's Hospital simplified diet charts, some unique and delicious recipes for the whole family featuring the veggie of the month - potatoes, and a thoughtful article on managing the PKU diet at daycare.

Over the next year, I look forward to reconnecting our Illinois community through surveys and polls to learn our challenges, gaps, successes and event feedback. Additionally, we encourage readers to submit questions or personal advice on navigating a PKU diagnosis throughout the life span. We hope this newsletter serves as a source of inspiration and collaboration for all those involved in our PKU and rare disease communities!

I would like to take a moment to thank my fellow board members and the entire fundraising committee for the Lifting the Limits for PKU Chicago hosted by the NPKUA. We have been working diligently since last year to put together a wonderful evening full of entertainment and inspiration to raise significant funds for the NPKUA programs and research in the hopes of achieving the ultimate goal - a cure for PKU. We invite all of our members, colleagues, and friends in the greater Chicagoland community to attend this one-time event or consider a monetary donation and make a difference in the lives of all those affected by PKU.

Sincerely,

Kirsten Frankfort & The PKU Organization of Illinois

★ *Special thank you!* ★
Soo Shim & Anne Kozek

UPCOMING EVENTS

NEW PARENT CAFE

Enjoy munching & mingling with other PKU families at the Naperville Alive Center. Learn tips and tricks to guide you in the early days of parenthood + PKU!

April 14 | presented by PKU Org of IL

LIFTING THE LIMITS FOR PKU CHICAGO

Please join us at the Lincoln Park Zoo Lion House for this one-time event! It will be an evening full of entertainment, inspiration, and fundraising for the ultimate goal - a cure for PKU!

May 4 | presented by NPKUA

FAMILY FUNDAY

Bring the whole family to Blackberry Farm and enjoy a picnic lunch while mingling and playing with fellow PKU families!

June 1 | presented by PKU Org of IL

ILLINOIS LOW PROTEIN FAMILY CAMP

Enjoy a beautiful weekend at Great Oaks Retreat Center full of fun events, low protein food, and making memories with fellow low protein campers!

August 9-11 | presented by PKU Org of IL

We want to hear from you!

- need help navigating a unique or challenging situation?
- looking to share your experiences or advice
- want to share your favorite PKU recipe?

email us: thepkupress@gmail.com

special event feature!

There are many ways you can support the Gala!

- Purchase a **sponsorship** your company or family name will appear on all publication and media related to the event and all sponsorship levels include tickets to the event [click here](#)
- Make a **monetary donation**, no amount is insignificant donate now [here](#)
- Donate an **auction item** with local restaurant or experience gift cards [here](#)
- **Attend gala** for a night of fun by purchasing tickets to the event [here](#)



You are invited to

Lifting the Limits for PKU Fundraising Gala
Saturday, May 4, 2024 / 6:00 pm - 11:00 pm
Lincoln Park Zoo Lion House / Chicago, IL
Attire: Black Tie Optional
Complimentary Valet Parking

Please join us for a night of awareness, inspiration, fundraising and celebration.

Cocktails / Premier Silent Auction / Dinner
Live Auction / Entertainment

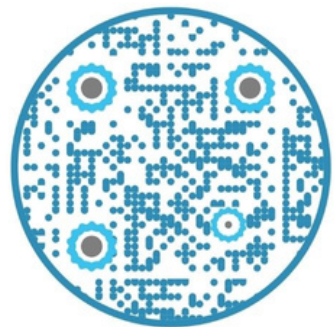
EMCEE:
Scott Pelley, award-winning journalist and
60 Minutes correspondent.

Contact Morgan Jurgens at Morgan@Npkua.org
for hotel room block booking options.

CHAIR:
 Grant Smith

HOST COMMITTEE:

Kim Endrizzi
 Kirsten Frankfort
 Kimberly and Mark Galen
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 Kristin Kopp
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 Brooke Kurkjian
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new
&
noteworthy



PKU IN THE NEWS

JNANA THERAPEUTICS: JNT-517

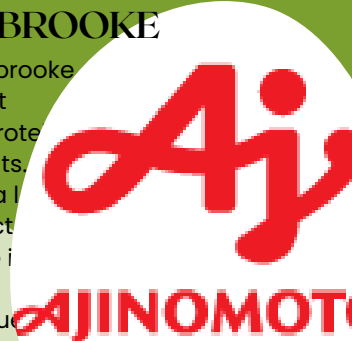
JNT-517 is a small molecule inhibitor of the Phenylalanine transporter SLC6A19 that works by blocking kidney reabsorption of Phenylalanine and excess phe is then excreted in the urine. It is a novel therapeutic drug taken orally by persons of any age or PKU phenotype. Phase 1A was a placebo-controlled, randomised trial that assessed the tolerability, safety, pharmacodynamics and pharmacokinetics of the Phe transporter SLC6A19 inhibitor JNT-517. Patients were dosed at various levels or given placebo. Phase 1A showed promising results, was safe and well tolerated with no adverse effects.

In August of 2023, the trial moved into Phase 1b to continue to assess pharmacokinetics, tolerability, and safety, as well as its effect on plasma and urinary Phe. Phase 1b is an ongoing, randomized, double-blind, placebo controlled study for people of ages 18-65. Interim results remain very positive - the drug was well tolerated, safe, and showed a 51% reduction in phe levels. The trial is currently enrolling for Phase 3.

WHAT'S (NOT) IN STORE?

AJINOMOTO CAMBROOKE

For over 2 decades, Cambrooke sought to create the most delicious, nutritious low protein food options for its patients. Recently there has been a buzz about lack of product availability. Cambrooke issued a statement in February describing the vendor issues, delays, closures, and discontinuation of products.



Cambrooke reassured its full commitment to its clients stating "We know change can be difficult, and each and every day we are looking for alternative solutions to ensure we deliver the best tasting and quality products we know you deserve and have loved for the last 20 plus years."

PHE-NOMENAL PEOPLE

Dr. Barbara K. Burton is a Professor of Pediatrics at the Northwestern University Feinberg School of Medicine who served for 20 years as Director of the PKU Clinic at the Ann & Robert H. Lurie Children's Hospital of Chicago. In 2019, she retired and is now focused on clinical research. Dr. Burton is an investigator in numerous natural history studies and clinical trials of new therapies for various metabolic disorders. She participated in the clinical trials leading to FDA approval of both sapropterin (Kuvan®) and pegvaliase (Palynziq®). She is currently an investigator in the Homology Medicines and Biomarin gene therapy trials in PKU. She has published over 200 peer-reviewed articles, 50 chapters in books and is an editor of two textbooks.

Dr. Burton is active in several professional organizations. In 2018, she received a Lifetime Achievement Award from March of Dimes and in 2019 she received the PKU Hero Award from the National PKU Alliance. Congratulations to Dr. Burton on an impressive, outstanding career! The PKU Community is eternally grateful for her expertise, leadership, and dedication to PKU!

“Taking care of patients with PKU and watching them grow and develop was one of the most gratifying aspects of my career. I always enjoy hearing from former patients and parents and keep up with how they are doing through the current treatment team at Lurie. It is such an exciting time with so many new developments in the treatment of PKU- I'm thrilled that I still have the opportunity to be involved on some level.”



VARY YOUR VEGGIES

OUR MONTHLY VEGGIE
SPOTLIGHT: POTATOES

Spud –tacular nutrition facts!

Did you know potatoes have more potassium than a banana? This versatile, varietal vegetable often gets a bad rap for its higher starch content and fried, processed, unhealthy versions. But it is a highly nutritious vegetable, providing a significant value in the daily recommended amounts of vitamins B6 and C, potassium and manganese, as well as being, naturally gluten free, and best of all its low pro!

EASY LOADED BAKED POTATO BAR

recipe by: Kirsten Frankfort

PREP TIME 15 minutes
COOK TIME 60 minutes
TOTAL TIME 75 minutes

Ingredients

russett potatoes
olive oil

toppings:

butter
sour cream
cheddar cheese (DF or non-DF)
steamed broccoli
white onion – diced small
green onions
tomatoes
eggplant “bacon” bits
jalapenos
salsa

**higher phe / non-
PKU options:**
chili
black beans
bacon bits

Instructions

1. preheat oven to 400 F
2. wash potatoes and leave to dry (both sides!)
3. use a fork to poke several holes on both sides of the potato
4. rub or brush both sides with olive oil and generously season with salt, pepper, and (optional) garlic powder
5. bake for about an hour or until soft through the center (depending on size of potatoes)
6. cut lengthwise and small slit crosswise and add a generous slice of butter
7. add toppings as desired - add them again! - and enjoy!

Notes

if serving family style - use a larger divided serving platter or lazy susan so young ones can independently load their potato!

adjust toppings depending on individual phe/protein allowance and dietary preference

THE SIMPLIFIED DIET

WHAT IT IS AND HOW TO USE IT

The simplified diet for PKU is a well-established alternative to the traditional measuring and weighing of fruits and vegetables in the low protein PKU diet. It evolved as a way to make the PKU diet more manageable, flexible, promote healthier options, and offer more palatable options than many medical foods.

Many clinics and dietitians have transitioned to the simplified diet for their patients by

- allowing for unlimited amounts of specific fruits and vegetables based on the nutritional content of having less than 75mg phenylalanine per 100 mg of food.
- reducing the patients overall tolerance by 30% to allow for the consumption of “free” foods.

The study, “Multiclinic Observations on the Simplified Diet in PKU”, concluded “While the approaches to implementing the diet vary slightly, all have observed that the Simplified Diet is easier to follow, encourages healthy food choices, and can improve the quality of life for patients with PKU as compared to the traditional counting method.”

Read more about the study

<https://www.hindawi.com/journals/jnme/2017/4083293/>

Simplified Diet Counted Foods

measure and count these foods

Artichokes	Mushrooms
Arugula	Orange Juice
Asparagus	Peas
Avocado	Potatoes
Bananas	Prunes
Broccoli	Prune Juice
Brussel Sprouts	Raisins
Cauliflower	Seaweed/Nori
Corn	Sundried tomatoes
Kale/Swiss Chard	Spinach
Mustard Greens	Sweet Potatoes/Yams

More information on counted foods

Low Fruits + Veggies (1 gm protein per serving)

- Prune juice, 1 cup
- Banana, 1 medium
- Broccoli/Cauliflower
- Kale
- Mushrooms
- Coconut, shredded
- Prunes

Serving sizes are 1/2 cup unless noted otherwise

Medium Fruits + Veggies (2 gm protein per serving)

- Corn
- Avocado
- Potato, 1 medium, baked
- Peas, frozen
- Spinach, cooked
- Sweet potato/yams
- Raisins

(Serving sizes are 1/2 cup unless noted otherwise)

Simplified Diet Uncounted Foods

no need to count these foods

<p>Vegetables</p> <ul style="list-style-type: none"> Acorn squash Butternut squash Cabbage Carrots Celery Chayote Squash Cucumber Green Beans Eggplant Jicama Leeks Lettuce (bibb, iceberg, romaine) Okra Onions Parsnips Peppers (all varieties) Pumpkin Radishes Rutabaga Sauerkraut Spaghetti Squash Summer Squash (zucchini and yellow) Tomatoes Turnips Yuca (cassava root) 	<p>Fruits</p> <ul style="list-style-type: none"> Apples - fresh and dry Apricots - fresh and dry Berries Cherries Cranberries - fresh and dry Fruit cocktail, canned Grapefruit Grapes Kiwi Lemons Limes Mango Melon (all varieties) Olives Papaya Peaches Pears Pineapple Plums Pomegranates Watermelon <p>Miscellaneous</p> <ul style="list-style-type: none"> Specialty low protein foods (<0.4 gm protein per serving) Herbs Sugary candy Butter/Ghee/Oils
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Always consult your dietitian and genetics team for specific diet instructions and allowances

TIPS *and* TRICKS

TO SPICE UP YOUR PKU TABLE!

FEATURED RECIPE:

LEMON GARLIC SMASHED POTATOES

recipe by: olivia adriance

Ingredients:

- 2 lbs small yellow or Yukon gold potatoes
- 4-5 lemons
- 8 cloves garlic, smashed
- parsley for garnish
- olive oil
- Salt & pepper

1. add potatoes to pot and cover with cold water
2. add juice of 2 lemons and smashed garlic
3. bring to a boil and cook until fork tender (~20 min)
4. drain and cool for 5 min (discard garlic)
5. preheat oven to 425
6. line a sheet pan with parchment paper and drizzle with olive oil
7. add potatoes and smash to ~1/2 in thickness
8. brush or spray with olive oil and season well with salt and pepper.
9. bake for 20 minutes - flip & add a slice of lemon to each potato
10. bake for 20-25 minutes until the edges are crispy
11. garnish with lemon zest and parsley
12. enjoy!!



air fryer

WHAT IS IT AND HOW TO USE IT

air fryers are similar to a convection oven by generating heat from a fan and circulating it around food

benefits

- reduces cooking time by ~50%
- healthier than deep fryer and microwave
- cooks a wide variety of food
- reheats pizza for a crispier crust in just 3-5 minutes

Favorite recipes to try!

- homemade fries or sweet potato fries (slice lengthwise, season with olive oil, salt & pepper, garlic powder)
- favorite "air fried" veggies - cauliflower, brussel sprouts, portabella mushrooms
- non-PKU'ers enjoy easy to cook fish, meat, and poultry in the air fryer as well!



Quick WEEKNIGHT DINNERS

CULINARY CORNER

MASTERING PKU COOKERY



EASY POTATO HASH

recipe by: Kirsten Frankfort

- potatoes
- sweet peppers (variety -green, yellow, red)
- onion
- garlic powder
- paprika
- olive oil

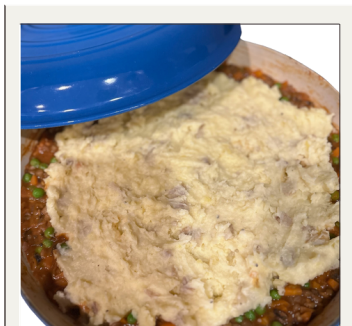
Toss all ingredients together. bake 400 ~30 minutes

Breakfast for dinner?!

- add Cambrooke "eggs"!
- wrap it all up in a low protein tortilla for a breakfast burrito!



Luck the Irish



VEGETARIAN SHEPHERDS PIE

recipe by: Budget Bytes

- 3 carrots (diced)
- 2 celery stalks (diced)
- 1/2 onion (diced)
- 2 cloves of garlic (minced)
- 8 oz mushrooms (baby portobello or button - diced)
- 1 cup peas
- 1 c cooked lentils (*optional or make separate)
- 4 cups mashed potatoes
- dried thyme
- paprika
- 1 tbsp tomato paste
- 1 tbsp flour
- 1 cup vegetable broth
- salt and pepper

Preheat oven to 400 F.

Sauté carrots, onions, and celery until soft. Add garlic then mushroom. and sauté until soft.

Add tomato paste and flour to the skillet - stir and cook until vegetables are coated and pastry mixture begins to coat the bottom of pan.

Add vegetable broth stirring to dissolve the flour and tomato paste. Bring to a boil and add peas and lentils Add seasonings

Transfer to oven safe dish or use pan. Top with mashed potatoes.

Bake the shepherds pie in the fully preheated oven for 15 minutes or until everything is heated through.



PHONE A FRIEND

Managing PKU diet at daycare

Written by: Soo Shim, LCSW & Anne Kozek, RD

Most daycare facilities are willing and capable of managing your child's PKU diet and treatment. Communication and planning are the most important aspects of ensuring that your daycare provider can manage your child's PKU. To help your daycare provider and ensure your child's safety, the following tips may be helpful:

- Explain PKU. It is important to be open with your daycare providers so that they know your child's needs. (See "Explaining PKU" at the beginning of this chapter.) NPKUA -New Parents Guide
- Provide lists. Make sure they are simple and easy to read. Outline foods allowed and not allowed in your child's diet. Stress the importance of drinking all the formula while at daycare.
- Offer to help communicate to staff. It may help your daycare provider if you come to the center to talk to staff about PKU. This can help make your child's needs known by all who may meet him or her.
- Be "on call." Tell your daycare providers that when in doubt about anything to do with your child's care, they should call you (MY PKU Binder page 39 from NPKUA website NPKUA > What is PKU > My PKU Binder)

need help navigating a unique or challenging situation? looking to share your experiences or advice?

email us: thepkupress@gmail.com