

Liver Health Quality Improvement Project

Spring Learning Session 2024



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Content

1. Introduction and Brief Overview of the Project
2. Liver Health Surveillance Testing: why, what, and when?
3. Lifestyle to Support Liver Health
4. Accessing Liver Health Education Materials



Liver Health



Exercise/Physical Activity



Emotional Health



Transplant



Birth Control & Pregnancy

Five Themes

Liver Health Project Team Members

ELT Sponsor: Carole Lannon

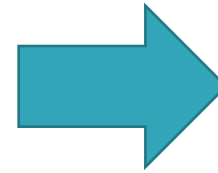
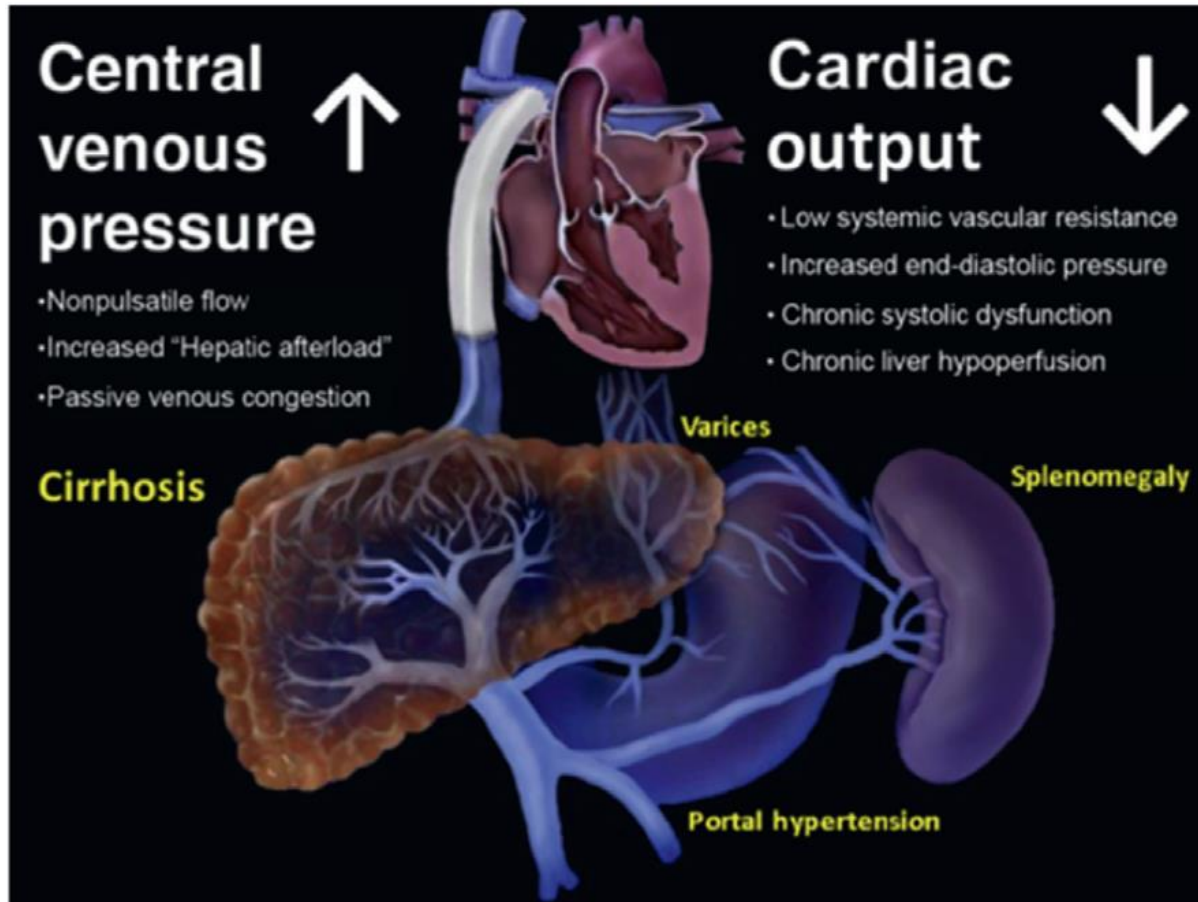
QI Workgroup Sponsor: Sal Ginde

Project Development Workgroup: Andrew Cheng, Jennifer Conway, Daniel Ganger, Alexis Gumm, Autumn Jones, Jana McClymonds, Cara Morin, Anita Pai, Mary Rouzer, Jack Rychik, Anita Saraf

Expert Feedback Workgroup: Letitia Aloysius, Shahnawaz Amdani, Adrian Dantzer, Noelle Ebel, Greg Fleming, Amber Hildreth, Taylor Houlihan, Katherin Madigan, Mandi McCormick, Nadia Naz, Melanie Parent, Elizabeth Rand, Suren Reddy, Rusty Rodriguez, Jose Silva Sepulveda, Rachael Suddarth, Divya Suthar, Milly Weisert, Kerry Wong

Network Support: Becky Collins, Adriana Ley-Chavez

Fontan-Associated Liver Disease (FALD)



Abnormal laboratory data

Progressive liver fibrosis

Cirrhosis

Liver nodules

Hepatocellular carcinoma

FALD Knowledge and Care Gaps



Although FALD is common and a clinically important issue after the Fontan operation, there are several gaps in knowledge:

1. Surveillance

No consensus on how to monitor progression of FALD → high degree of variability among centers

2. Education

Variation in FALD education and counseling for patients and families

3. Management

Cardiac centers may vary in access to and collaboration with hepatologists (liver specialists) with knowledge/experience with FALD

Global Aim

Optimize the liver health, quality of life and longevity of individuals living with Fontan circulation



Key Drivers to Address



Ensure reliable assessment and surveillance of FALD at Care Centers



Promote consistent education of patients/families and clinicians about FALD



Consider multidisciplinary care models with dedicated hepatologists to address FALD

FON Liver Health Project

Interventions

Global Aim

Optimize the liver health, quality of life and longevity of individuals living with Fontan circulation

Project Aim

Reliable clinical management of the liver health of individuals living with Fontan circulation as measured by (TBD)

Population

All individuals living with Fontan circulation, families, and all clinicians who provide care; not limited to patients enrolled in registry

Key Drivers

Ensure reliable assessment and surveillance of FALD at Care Centers

Promote consistent education of patients/families and clinicians about FALD

Consider multidisciplinary care models with dedicated hepatologists to address FALD

Recognize and support the use of evidence-informed strategies to support LH surveillance

Incorporate patient preferences, risk factors, & constraints into LH surveillance strategies

Engage FON centers to apply and disseminate the LH surveillance strategies developed

Develop & deliver materials leveraging existing resources for care centers to provide necessary information/education to clinicians and patients/families

Promote dissemination of information/education to the larger community (to patient/family groups or to centers without specialized providers)

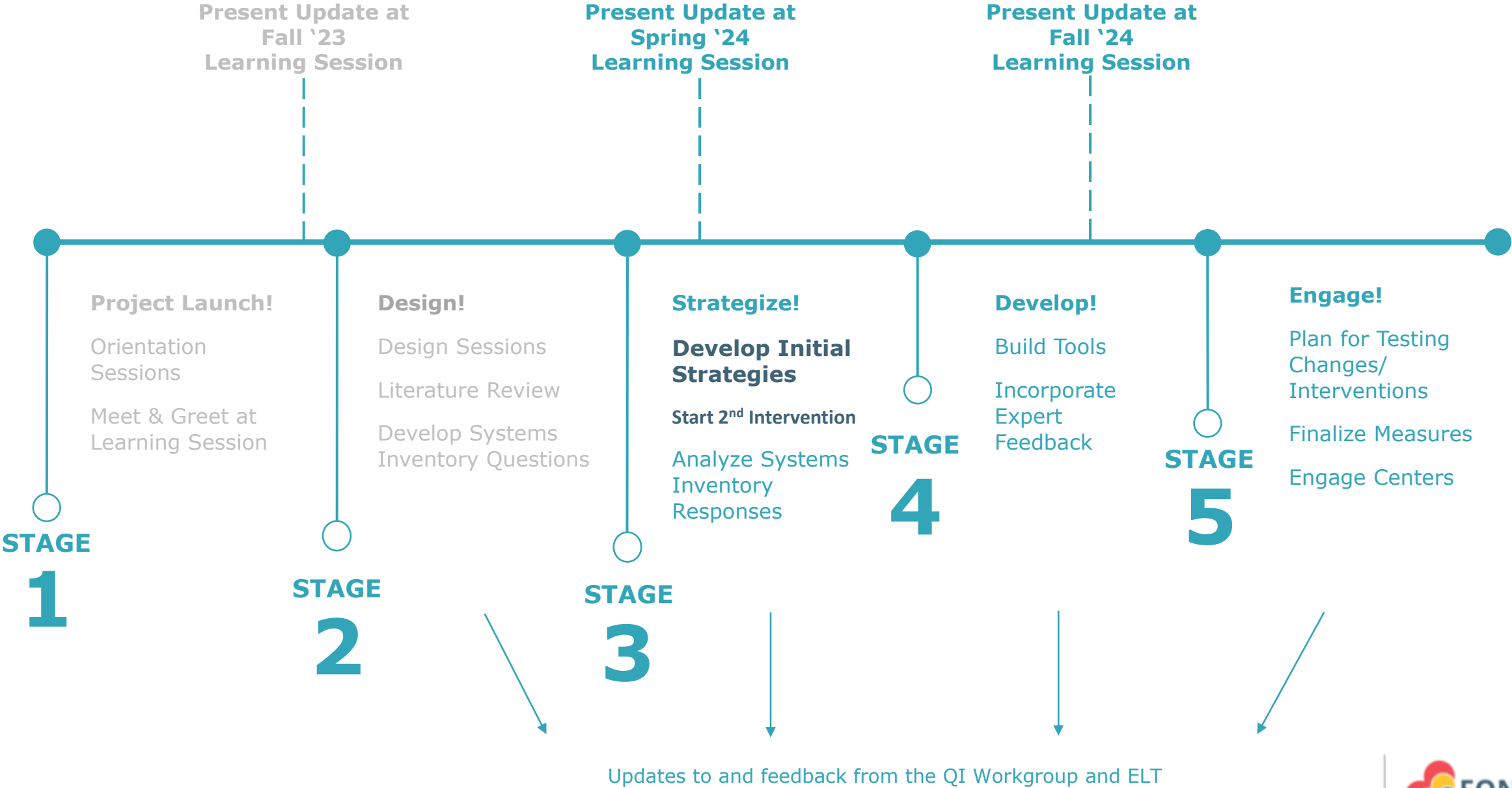
Develop LH guidance and education materials to support patients in the transfer to adult care

Develop guidance & education materials to support community GI providers and hepatologists

Develop & disseminate strategies for care centers to develop collaborations with GI providers

Build a community platform (message board) for providers to consult each other on LH concerns

Liver Health Project Timeline



First Intervention in Development

FALD Education for Patients and Family Members



Patient Experience: Not Receiving Timely Liver Health Information

Presented by Mary Rouzer DNP, APRN, FNP-C

Lurie Children's Hospital/Northwestern Medicine



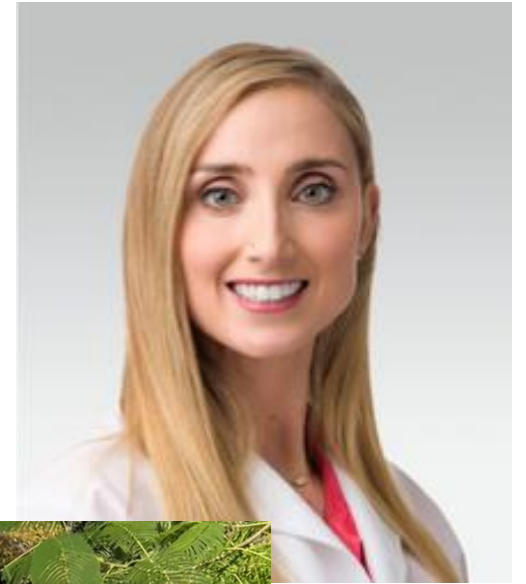
What do you most want to know regarding LH testing?



- 1. Why is liver health testing needed**
- 2. What age to start screening**
- 3. Which tests are preferred**
- 4. How often do we need testing**
- 5. Other: Please type it in the chat!**

Introduction

- Hepatology Nurse Practitioner
 - CHD Hx: Double inlet hypoplastic left ventricle with transposition of the great vessels, coarctation of the aorta
 - Status post Fontan ('94)
 - Cardiac care center: Private Practice/Lurie
 - Hepatology care: Northwestern Medicine
-
- Picture with my daughter (Luna) and husband (Nick)





Sensitive Material



The content that is about to be shared might be considered sensitive for some members of the community.

Please feel free to take a break or step away, if needed.

Liver Health: Patient Experience

- No guidance on when or why to see a hepatologist
- Limited information of the impact of the heart on the liver and why establishing care with a liver doctor would be necessary
- Saw a general gastroenterology that was interested in hepatology after post operative ascites following a cholecystectomy – EGD, imaging, paracentesis was completed.
- No additional f/u with any GI or hepatologist until I started working at NM
- When asking cardiology re: liver health I was assured they would follow my liver health and refer me when necessary

Liver Health Surveillance

Why is testing necessary?

- Fontan circulation causes blood to accumulate in the liver leading to liver swelling.
- With time, this swelling can lead to harm and progressive scarring of the liver.
- Usually there are no symptoms and you will feel well, but your doctor will do surveillance to identify any of the following: liver dysfunction, development of large veins in the throat and stomach that can bleed, enlarged spleen, fluid in the abdomen, and although rarely, liver cancer.

When to start testing:

- About 10 years after Fontan surgery, or sooner if your cardiology provider indicates
- Lifelong testing will be done at regular intervals determined by your healthcare professional.
- Testing may include imaging (such as ultrasound, CT or MRI), blood work, and/or a liver biopsy.

Who will order the tests?

- Your cardiologist and/or a liver specialist (hepatologist) familiar with Fontan patients
- Ask your provider what your personal surveillance plan should be

Blood Tests

- These tests show whether your liver is damaged and how well it is working.
- How often your doctors order blood tests depends on your age, risk factors for liver disease, and prior test results.

Test	Possible meaning of test if abnormal				
	Liver cells are inflamed	Bile is not draining from liver well	Liver is not working well	Blood is not flowing into liver well (portal hypertension)	Liver cancer might be forming
Alanine and aspartate aminotransferase (ALT and AST)	X				
Total bilirubin		X			
Alkaline phosphatase	X	X			
Gamma glutamyl transferase (GGT)	X	X			
Albumin			X		
Prothrombin time and international normalized ratio (PT and INR)			X		
Platelet count				X	
Alpha fetoprotein					X

Liver Imaging

- These tests involve taking pictures of your liver and other organs by ultrasound, computed tomography (CT), or magnetic resonance imaging (MRI). Each type of imaging test has different ways to show scarring or damage of your liver and identify liver nodules.
- The results of these tests don't always match the results of liver biopsy.
- How often your doctors order imaging depends on your age, risk factors for liver disease, and prior test results.

Imaging tests

Ultrasound (US)

- Uses sound waves to create pictures of your liver and sometimes other organs, such as your spleen
- Shows changes related to scarring and can find abnormal liver nodules, usually if they are bigger than 1 cm
- Sometimes uses a special type of intravenous (IV) contrast made of bubbles to help show liver nodules

Computed Tomography (CT)

- Uses x-rays to obtain pictures of your liver and other organs that may be affected, such as your spleen
- Shows changes related to scarring in your liver
- Provides detailed pictures of any abnormal liver nodules to show if they are benign or worrisome
- Requires the use of intravenous (IV) contrast, a clear liquid, to help show the liver tissue and liver nodules
- Safe for patients with devices or metal who cannot get an MRI

Magnetic Resonance Imaging (MRI)

- Uses a strong magnet to take detailed pictures of your liver and other organs that may be affected, such as your spleen
- Shows changes related to scarring in your liver
- Provides detailed pictures of any abnormal liver nodules to show if they are benign or worrisome
- Usually requires the use of intravenous (IV) contrast, a clear liquid, to help show the liver tissue and liver nodules
- Not safe for people with certain types of medical devices or metal in their bodies

Elastography

- A special test that detects the stiffness of your liver, which can help diagnose swelling, scarring, or other liver damage in your liver tissue
- Can be performed by US or MRI

Additional Tests

Liver Biopsy

- This test involves a doctor using a needle to obtain a very small piece of liver tissue. It can show how much scarring is in your liver and whether an abnormal nodule is cancer.
- This test only checks a very small part of your liver. It also does not show how well your liver is working.
- Since this test has some risks (such as bleeding), it is not done often. Some doctors get a biopsy at set times (such as every 10 years). Others only get a biopsy if they are concerned about the results of blood or imaging tests.

Endoscopy or Esophagogastroduodenoscopy (EGD)

- If your other tests suggest that you have severe liver disease, your doctor may recommend this test to look for swollen veins (varices) in your throat, stomach, and intestines.
- Knowing whether you have swollen veins (varices) is important because if they tear they can cause dangerous amounts of bleeding.

Limitations

- Sometimes the results of blood tests, imaging, and biopsy are not exactly the same

Patient FALD Perspective

My Journey with HLHS, FALD, and Teen Advocate -> Future Pediatric Cardiologist, I'm 17 years old.

By: Jana McClymonds



What do you most want to know regarding lifestyle to support LH?



- 1. How much alcohol is OK**
- 2. What can I do to improve my liver health (exercise, diet)**
- 3. What food should I eat more of, and what should I avoid? Supplement use?**
- 4. Other: Please type it in the chat!**

About Me

- My life has been a rollercoaster right from the very beginning. I was born with a little *surprise package - HLHS*.
- Norwood, Bi-Directional Glenn, and Fontan and post Fontan fenestration.
- Diagnosed with FALD at 15, the temporary medical team seen (Covid era), told me they didn't want to scare me but things like this looked like cancer, but not cancer, they weren't sure!
- My parents asked; "How common is this in HLHS?" *Their answer "Not often, they never seen anything like this and needed to figure it out". WOW*
- The wait began. The hematologist gave me a list of things I could not eat or drink, medicine I could not take, the medical team was very inconsistent.
- I turned to social media to look for other HLHS patients with FALD.

- *More research is needed*
- *I have so many questions*
- *Who do I trust?*

Compression Socks & Oxygen Needs

Traveling (Flights)

- For my 17th birthday, I flew halfway around the world to the UAE and the Maldives in the Indian Ocean. I snorkled 12 miles out of the atoll with turtles – did not do scuba diving.
- I wore compression socks for most of the flight and wore my oxygen concentrator when sat's dropped to 92. Any flight over 4 hours, I ensure compression socks and an oxygen concentrator.

High Altitudes

- Above Park City, Utah, 9000+ elevation, I did sleigh rides for Christmas – I did not wear compression socks nor used oxygen. I visited for less than 3 hours.

24 Hour Marathon

- Every year, I walk 24 hours with my 82-year-old grandfather, retired Sgt. Major of the Marines, raising money for ROTC. This year, I walked 16 miles and wore compression socks.



Alcohol, Meds, Caffeine, & Supplements

Alcohol & Caffeine

- In the Maldives for my 17th birthday, I drank Champagne. After a few sips I was lightheaded and very happy. I take aspirin daily. College is in a few months - alcohol will be an experience I'm sure at 21.
- I drink unsweetened iced tea (30-60mg), green iced tea (60mg), and cherry coke (30mg), I pay attention to caffeine. No energy drinks, no coffee.

Food

- Love salad, fruit, a good steak, veggies, nuts/seeds, and croissants. On occasion, ice cream, cupcakes, and candy.

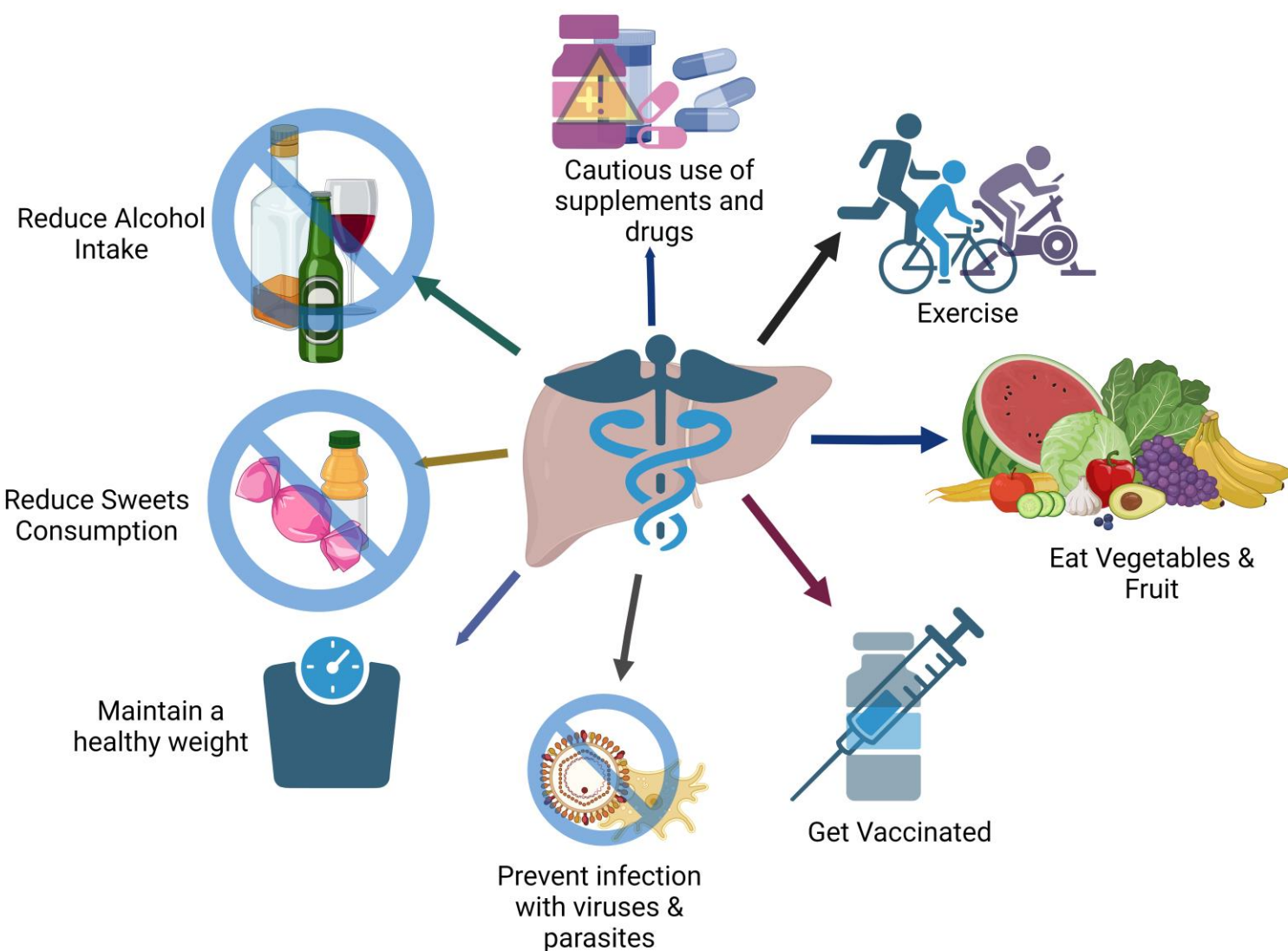
Medicine & Supplements

- Multi-vitamins, vitamin D, C and *question* immunity *N-Acetyl-L-Cysteine 600mg, and Quercetin Dihydrate 250mg*
- Medicine (Prescription)
 - Wisdom Teeth: Meds for sedation? Pain meds? I have questions.
 - Anxiety? Can I have valium? / Tension Headaches: Can I take migraine meds – like Fiorciet?
 - Birth Control? While it is not on my mind now, what can I take when I am older?

Education on lifestyle to support liver health

Draft – In Development

DRAFT



DRAFT

What a parent of a young child needs to know

Presented by Autumn Jones, RN, MSN, FNP-C, and proud heart mom of Landon



(pic of us on his 3rd grade class field trip)



We want to hear from you!



Please type your response in the chat



Patients and Family Members:

- What would be most helpful to have access to relevant information?
- What formats (videos, handouts) would you want to receive?



Providers:

- What strategies have worked well to consistently deliver education about liver health to patients and families at your clinic?
- What barriers do you face to do this?

Background

- FNP for a convenient care clinic for the last 11 years
- Son (Landon) born with multiple critical CHDs in June of 2014:
 - *Shone's Complex with HLHS*
 - *Post Fontan since January 2017*
 - *Also post Pacemaker*
 - *Care Center: UPMC Children's Hospital of Pittsburgh*
 - *Started seeing hepatologist about 6 years post Fontan*



[Pics: Landon playing soccer; Selfie with my 3 kids (Little sister/Scarlett, Big brother/Camden, & Landon); Landon enjoying a smoothie; Family selfie including Hubb/Vinny]

Education on what to expect after the Fontan

- At first it's all about survival/making it through the surgeries (the 1st few years are focused on that).
- After the Fontan, you have a bit of room to breathe, things can calm down a bit.
- That's when I first learned of how the liver is impacted by the Fontan circulation (so my son was about 3 years old when I 1st learned this). For whatever reason, this information completely shocked me.
- *A part of me wishes I had been aware of this issue sooner, but I realize that I had so much other information and worries to focus on.
- Already, by age 6 he had elevated liver labs and at age 8 he started seeing a hepatologist through our Cardiac Care Center.
- *I am so thankful for how well Landon does overall, but I want there to be more that can be done to prevent liver complications post Fontan (that's why I became involved with this organization).
- [One of the things I am focused on is what is the best way for parents to access liver health info.](#)

Delivery Methods to provide education to patients and family members

1. Short videos on social media or FON website

Examples

- Video displaying information with links to more detail
- Educational videos explaining why liver is affected by Fontan circulation
- A cardiologist and hepatologist discussing liver health, showing collaboration among specialties
- Patients discussing their experiences with liver health
- Scripted clinic encounters w/ patients being counseled about liver health (i.e. alcohol, lifestyle, etc)
- Cartoons talking to each other providing information

2. Printed handouts or pamphlets at clinic visits or on FON website

3. Information on After Visit Summary (AVS) and/or patient portal (i.e. EPIC MyChart)

Early Look: 1-page Handout



DRAFT

About Liver Health Surveillance

Why is surveillance or testing necessary?

Fontan circulation causes blood to accumulate in the liver leading to liver swelling. With time, this swelling can lead to harm and progressive scarring of the liver. Usually there are no symptoms and you will feel well, but your doctor will do surveillance or testing to identify any of the following: liver dysfunction, development of large veins in the throat and stomach that can bleed, enlarged spleens, fluid in the abdomen, and although rare, liver cancer.

When does testing start?

About 10 years after Fontan surgery, or sooner if your cardiology provider indicates. Lifelong testing will be done at regular intervals determined by your healthcare professional.

Who orders the tests?

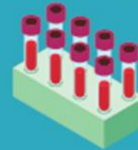
Your cardiologist and/or a liver specialist (hepatologist) familiar with Fontan circulation. Ask your provider what your personal surveillance plan should be.

How often does testing occur?

How often your doctors order imaging depends on your age, risk factors for liver disease, and prior test results. Repeating these tests is needed to monitor for liver damage and liver nodules. A young child might have imaging rarely, while an adult with risk factors might have testing more than once per year.

Types of Liver Testing

Blood Tests



These tests show whether your liver is damaged and how well it is working. Since each test provides slightly different information, your doctors will usually order many tests at once.

Blood tests cannot show how much scar tissue is in your liver or if there are abnormal nodules in your liver.

Liver Imaging



Liver imaging involves taking pictures of your liver and other organs by ultrasound, computed tomography (CT), or magnetic resonance imaging (MRI). Each type of imaging test has different ways to show scarring or damage of your liver and identify liver nodules.

The results of these tests don't always match the results of liver biopsy, which is an additional test that your doctor may order.

How Can I Improve My Liver Health?

Other liver diseases: Other liver diseases can increase your risk of accelerating liver damage. This includes fatty liver disease, diabetes, alcoholic liver diseases, etc. For a complete list [here](#).

Avoid toxins: Some herbal supplements, over-the-counter medications, and prescription medications can damage your liver. Please check with your doctor or livertox.nih.gov to check for potential liver toxicity with any new medications.

Limit alcohol intake: Excessive alcohol use can cause liver damage. Talk to your provider about how much alcohol is safe for you.

Reduce Alcohol Intake

Reduce Sweets Consumption

Healthy weight: Ideally maintain a BMI of 18 - 25 kg/m²

Maintain a healthy weight

Prevent infection with viruses &

Cautious use of supplements and drugs

Exercise

Eat Vegetables & Fruit

Adopt a healthy diet: Eat a balanced diet with foods from all food groups, including fruits, vegetables, grains, meat and beans, milk, and oil. Avoid foods with added sugars such as soda, mixed drinks, pastries, and sweets.

Protect yourself against infections: Hepatitis B and C can infect and damage the liver. Hepatitis occurs from infected needles, including tattoo and piercings, and infected body fluids. Hepatitis vaccines can prevent

Get Vaccinated

Feedback?
Questions?

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Feedback? Questions?

