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Issue 11



SVR Extension Newsletter

WELCOME!

In this issue of the SVR newsletter we have included some exciting findings from the Pediatric Heart Network's FUEL trial, and also, a brief summary of some new work related to leg strength for patients who have had a Fontan. If you have any questions about either of these articles, please don't hesitate to talk with your pediatric cardiologist or with the SVR research team at your site. We are also happy to share with you an article written by Brenda Henne, an educator and school liaison, describing how neurodevelopmental evaluations can be a valuable tool in getting needed educational assistance. We hope that this information is helpful to you.

We are so delighted to share some beautiful artwork here from our SVR participants as well! Thank you to all of our artists who have submitted their pieces so far. We will look forward to sharing more art work in future newsletters. Definitely talk with your research team if you are interested in submitting art for a future issue.

As we finalize this newsletter, we have found ourselves, and the whole world, in unusual times due to the COVID-19 pandemic. We are hoping that each of you and your families are doing well despite issues with the pandemic. While we continue to learn more each day, so far we are happy to see that the COVID-19 virus is only rarely having direct health effects on children. Nonetheless, these are stressful times. We have included in this newsletter some helpful tips for managing stress for children and for parents, compiled by a group of psychologists from the Cardiac Neurodevelopmental Outcomes Collaborative (CNOC).

We know that some of you were planning to come in for evaluations related to this study in the Spring and Summer 2020 and at this point there may still be uncertainty at your site about when in-person visits for research participation will be possible. Information on next steps will be shared by the research team with you as we learn more.

Thanks again to each of you for your ongoing engagement in the SVR studies and for your help in identifying the best treatments for children with hypoplastic left heart syndrome and other related congenital heart diseases.

Please take care.

Sincerely,

Caren Goldberg, MD, MS
SVR III Study Chair
Professor, Pediatric Cardiology
University of Michigan



Pediatric Heart Network (PHN)

"Thanks again to each of you for your ongoing engagement in the SVR studies and for your help in identifying the best treatments for children with hypoplastic left heart syndrome and other related congenital heart diseases."

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PHN FUEL TRIAL: COMPLETED AND PUBLISHED

The Pediatric Heart Network recently completed and published the results of the Fontan Udenafil Exercise Longitudinal (FUEL) Trial. This trial included 400 adolescents (12-18 years of age) from 30 sites around the world who were born with single ventricle heart disease and surgically palliated with the Fontan procedure. Participants at each site were given either udenafil, a medication that can lower pulmonary pressures, or a placebo (sugar pill) for six months. At the beginning and end of the trial participants underwent standardized exercise testing. Those who received udenafil demonstrated improvement in moderate levels of exercise compared to those who received the placebo. At peak exercise, there was a suggestion of improvement in those who received udenafil, but this improvement did not reach statistical significance. Udenafil was well tolerated with



side effects limited to those known to be associated with this type of medication. The Pediatric Heart Network is currently engaged in further evaluations to understand if there is an exercise benefit for adolescents with a Fontan when udenafil is given for a year or more. The results from the first part of this study were published in full in *Circulation* in November 2019. If you are interested in learning more about this study and the findings you can talk with the Pediatric Heart Network team at your site as well.

David J. Goldberg, MD
Associate Professor of Pediatrics
The Children's Hospital of Philadelphia

“Those who received udenafil demonstrated improvement in moderate levels of exercise compared to those who received the placebo.”

SINGLE VENTRICLE FONTAN PHYSIOLOGY

Over the past 5 decades, outcomes have improved dramatically for patients with single ventricle Fontan physiology. We are thrilled to see so many patients growing, learning, and thriving! Many patients find that regular exercise is important to their wellbeing. Regular exercise may help patients stay healthy and feel happier and more satisfied.

We are learning more about exercise in Fontan patients and ways to help patients exercise better. The heart, lungs, blood vessels, and muscles all affect exercise performance. PHN investigators previously showed that heart function accounts for a lot of the differences in exercise ability between Fontan patients, but other factors are important as well. Our group is interested in studying the skeletal muscles – particularly the muscles in the legs – in Fontan patients. The leg muscles help to increase blood flow in the Fontan circulation, especially during exercise. Building muscle mass in the legs through an exercise training program may result in improved exercise ability.

Our research has shown that Fontan patients often have less muscle in the legs compared to non-Fontan patients. However, Fontan patients with more muscle in the legs performed better on exercise stress test and exercise cardiac MRI. In similar work, Dr. Rachael Cordina's group in Australia showed that muscle mass and exercise performance were improved in adult Fontan patients after participating in a resistance exercise training program. We are hopeful that exercise training can also help build muscle mass and improve exercise performance in pediatric Fontan patients and we are now studying this.

With the support of the PHN Scholars program, we are currently performing a dual-centered study at the Children's Hospital of Philadelphia and Cincinnati Children's Hospital Medical Center to test the effects of a 6-month exercise program on muscle mass and clinical status in adolescent Fontan patients. The intervention includes in-person sessions and at-home exercises. We are studying the effect of the training on muscles, peripheral blood vessels, quality of life, and performance on bike stress test and exercise cardiac MRI. We are very excited about this pilot study and hope to gain more data for a larger trial and to eventually improve exercise ability and other outcomes for Fontan patients. We are very grateful to all the study participants!

Catherine Avitabile, MD

PHN Scholars Grant Recipient

Children's Hospital of Philadelphia

Assistant Professor of Pediatrics

University of Pennsylvania School of Medicine

*“WE ARE LEARNING MORE ABOUT EXERCISE IN FONTAN PATIENTS
AND WAYS TO HELP PATIENTS EXERCISE BETTER.”*

NEURODEVELOPMENTAL ASSESSMENTS AND FOLLOW-UP – WHAT’S THIS?

My previous article, reviewed the strength of planning, partnerships and communication as tools that build solid educational foundations. It ended with the following charge for all of us:

*“**Purposeful planning**, along with strong **partnerships** resulting in **communication** that is clear, concise and accurate, reduces potential barriers to the school experience for our children leading to better quality of life outcomes . . . educationally, socially, and physically.*

This collaboration - sharing of information in a format that is readily understood by the school - increases the school’s knowledge; thereby, reducing potential barriers to the helpful supports and services that our children require. Yes, our children’s educational needs fall somewhere on a continuum, but the planning that is required must be predictable and consistent.”

Information. This article continues with this charge . . . Beginning when our children are very young, and throughout their developmental years, obtaining information about their developmental and learning strengths is a priority. Information helps us identify areas of strength and need, and the interventions, supports and/or strategies that will build solid foundations for later learning and future development. We obtain this information through a comprehensive neurodevelopmental assessment. A comprehensive assessment of children with a wide range of developmental, behavioral, emotional, and learning challenges. Obtaining information, identifying plans for supports and resources, and sharing the information with stakeholders is the goal of a neurodevelopment assessment and the subsequent follow-up conversations that occur along the way.

Need. With the advancements of medical procedures and treatment, children born with congenital heart defects (CHD) have opportunities for a longer life . . . and greater opportunities for a wonderful quality of life. As partners, we can make a significant impact on a child’s quality of life during specific milestones: infancy, toddlerhood, early childhood, and their school age years. **There is overwhelming evidence that children who undergo cardiac surgery during their first years of life are at a higher risk for developmental, learning, and/or behavioral challenges.** These risks may be present early or appear along the way as a child’s brain continues to develop. This continued development encourages us to have our child assessed for challenges at specific developmental milestones. After an initial assessment in infancy, these milestones are at age 3, age 5, and then every three years thereafter.

Identification. Neurodevelopmental assessments help our children’s families with early identification of a wide range of developmental, behavioral, and/or learning challenges. These challenges may include:

- Motor skills, both fine and gross
- Speech and language use
- Feeding problems
- Attentional, behavioral, and emotional
- Social skills - getting along with friends, family, and class mates
- School readiness in the areas of math, reading and writing
- Executive functioning – such as time management, organization, planning, initiating

Planning. When the results of the neurodevelopment assessment indicate an area(s) of challenge, specific interventions, strategies, and support can be implemented to address the area(s) and enhance further learning and development. The earlier an area(s) is identified, and the support is implemented, the greater the opportunity for improved learning and developmental outcomes. We want to identify challenges early, create effective learning plans, and implement these plans as soon as possible. This is why early identification and the information provided by the neurodevelopmental assessment becomes so important.

Partnership. Providing our children with opportunities for the very best outcomes begins with knowledge. The neurodevelopment assessment is a strong and comprehensive tool that provides us with our child’s developmental strengths and learning needs. Having ongoing and in-depth knowledge to share with developmental and learning specialists greatly enhances our children’s outcomes.

Communication. Obtaining information, identifying plans for supports and resources, and sharing the information with stakeholders is the goal of a neurodevelopment assessment and the subsequent follow-up conversations that occur along the way.

Neurodevelopmental Assessment and Follow-up: Information. Need. Identification. Planning. Partnership.

Brenda J. Henne
School Liaison/Educational Specialist
Congenital Heart Center
C. S. Mott Children’s Hospital | Michigan Medicine | University of Michigan



Coping with Stress and Worry during COVID-19:

Information for People with Congenital Heart Disease and their Families

Many people are feeling stressed and worried about the coronavirus disease 2019 (COVID-19). This is natural. These feelings can be even stronger for people with congenital heart disease (CHD) and their family members. When we don't know much about something, like this virus, we can feel unsafe. But, remember, people with CHD and their families have been through difficult times before and have already learned some ways to handle stress that can be helpful now.

The Cardiac Neurodevelopmental Outcome Collaborative (CNOC) put together this information to help people with CHD and their family members handle stress and worry that you may have about COVID-19. Remember, we all handle challenging situations differently, and it's important to do what feels right to you.

Take breaks from news and social media. News about COVID-19 is available day and night on the news and social media. You may feel like you need to keep watching, reading or listening to these updates to stay on top of what is happening. But if you spend too much time on COVID-19 updates, you can get even more worried and stressed. Instead, think about checking the news and social media for only a short time a couple of times a day. That way you still know what the federal/state public health and safety experts are suggesting. We recommend not getting updates in the hour before you plan to go to sleep.

Take care of your body. It is always important to take care of your body, but this is especially important during times of stress. Try to get to sleep early and eat healthy meals. Walk outdoors when it's less crowded outside or find free exercise videos online. The body and mind are connected, and taking care of your body will help you handle stress and worry.

Deep breathing and mindfulness exercises can help. Just breathing slowly and deeply for a few moments can help you relax. You can do this as many times as you need to during the day. You can learn about other relaxing exercises using apps on your phone or on the Internet. Many only take a few minutes to do. Some apps have free breathing, mindfulness, and guided meditation exercises (<https://www.headspace.com/covid-19>, <https://blog.calm.com/take-a-deep-breath>). Write down other things you like to do that relax you and try to do at least one of these every day.

Try to plan out your day and make a list of goals that you can finish each day. These goals can be for your work, family, home, or hobbies. Even if you are stuck at home, try to maintain your morning and evening habits (meals, showering, getting dressed, etc). If you have a plan and goals you can finish each day, you will feel more on top of things during this hard time.

Coping with Stress and Worry during COVID-19:

Information for People with Congenital Heart Disease and their Families (Contd.)

Connect with people who are kind and caring using your phone, tablet or computer. You might enjoy connecting with other people affected by CHD (patients, parents, etc.) because they have had experiences like yours. Social media connections can help you feel less alone. But remember that people on social media may be scared too and may talk about the worst possible situation. If you start getting more worried or scared by what people are saying, then you may want to stop for a while so you don't get more upset.

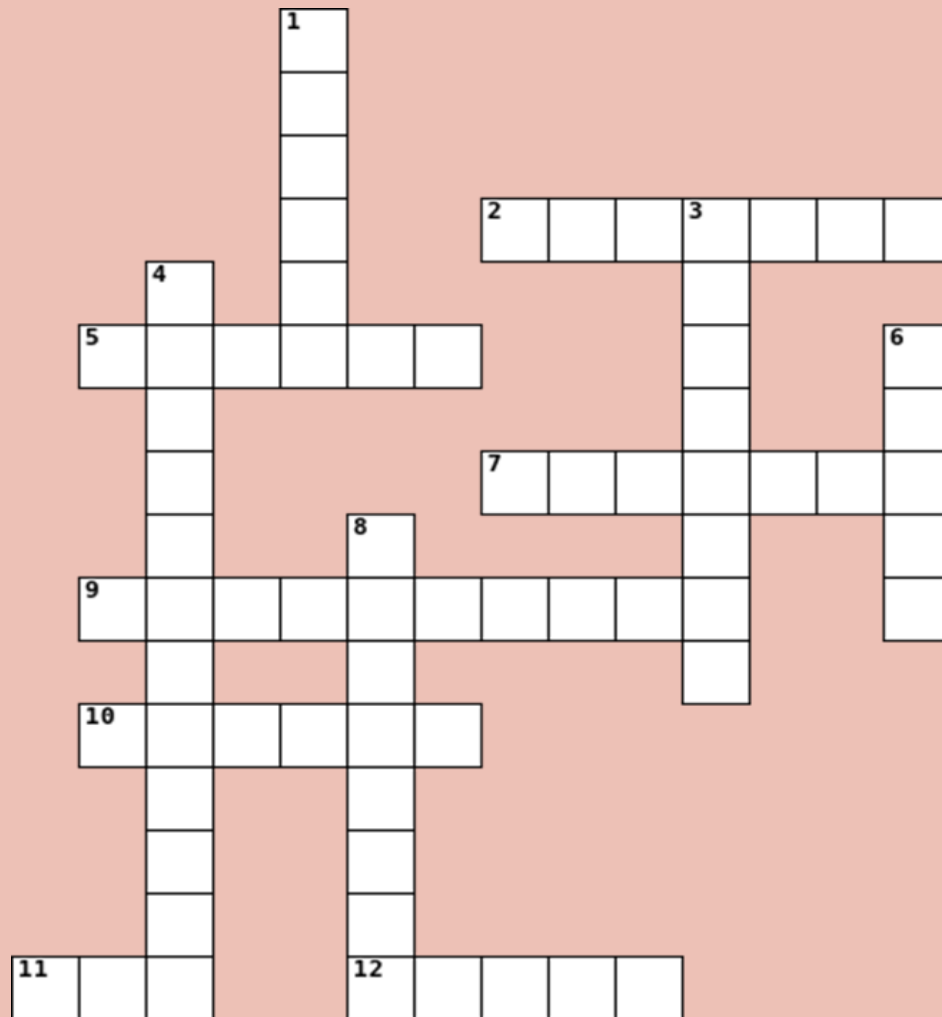
Talk to a mental health professional for help. You may be able to speak to a mental health professional (psychologist, social worker, therapist) using online video meetings, without having to leave the house. This is called telehealth. If you feel like your sadness or worries are making it hard for you to get through your day, speaking to a mental health professional might help you. (For example, you might feel really scared often or cry a lot. You could have thoughts or pictures in your head that don't go away or find it hard to take care of yourself, others or your work tasks. You could have trouble sleeping or not feeling hungry like you used to.) To find a mental health professional, contact your insurance company for a list of mental health clinics that take your insurance. Call the clinics to find out if they provide therapy using telehealth. You can also call your primary care doctor or your cardiologist or heart doctor's office for suggestions of where to call. Many hospitals have social workers and psychologists on staff who may be available for telehealth or may be able to help you locate someone who provides telehealth services.



The links below have information on how to deal with COVID-19 related worry and stress:

- CDC: <https://www.cdc.gov/coronavirus/2019-ncov/prepare/managing-stress-anxiety.html>
- SAMHSA: <https://store.samhsa.gov/product/Taking-Care-of-Your-Behavioral-Health-During-an-Infectious-Disease-Outbreak/sma14-4894>
- American Psychological Association: <https://www.apa.org/practice/programs/dmhi/researchinformation/social-distancing>
- Centre for Addiction and Mental Health: <https://www.camh.ca/en/health-info/mental-health-andcovid-19>

PHN CROSSWORD PUZZLE



Across

2. people from around the world compete in different sports at the _____ games
5. MRI uses a big one of these to take pictures of your heart
7. a sea animal that uses ultrasound (just like an echocardiogram)
9. another word for 'born with'
10. Harry_____, a wizard who attends Hogwarts
11. a flying animal that uses ultrasound
12. the organ that pumps blood to the rest of your body

Down

1. what red blood cells carry from the heart to the body
3. a board game where you 'pass go' and buy properties
4. the doctor I see for my heart
6. the major organ used for breathing
8. how medical professionals learn the best treatments for patients

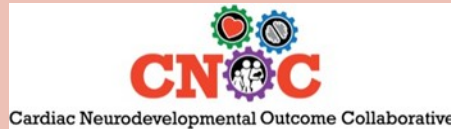
PHN ART GALLERY



-Alyssa



-Tess



Helping Children Cope with Stress and Worry during COVID-19: Information for Families of Children with Congenital Heart Disease

Many children and teens are feeling stressed and worried about the coronavirus disease 2019 (COVID-19). This is natural. These feelings can be even stronger for children and teens with congenital heart disease (CHD) and their siblings. When we don't know much about something, like this virus, we can feel unsafe. However, there are things that parents and family members can do to help children and teens handle these feelings in a healthy way.

The Cardiac Neurodevelopmental Outcome Collaborative (CNOC) put together this information for families of children with CHD. Remember, we all handle challenging situations differently and it's important to do what feels right for your family.

Try to remain calm and reassuring when talking with children and teens. Children and teens are looking to adults for information on how to react to COVID-19. Remind children that adults are working to keep them safe and teach them what they can do to help (washing their hands, keeping distance from others outside of the household). Try to limit adult conversations and media (television, radio) about COVID-19 around children.

Make yourself available for questions about COVID-19 and follow your child's lead. Your child or teen is likely to have questions about COVID-19 and may ask these questions at times when you are in the middle of other things. Try to follow your child's lead and, if possible, take a break from what you are doing to answer your child's questions. Be honest and give accurate, developmentally appropriate information. It is better for information about COVID-19 to come from trusted adults instead of social media or your child's friends. Some children may not have questions or concerns and that is okay too. Every child is different and will handle this situation in different ways.

Keep a routine and set limits on behavior. Predictable routines and limits on behavior will help children feel safe. Even though your child or teen may not be leaving the house, encourage them to continue their morning and bedtime routines (meals, showering, getting dressed, etc). It is also important for children and teens to know that adults are still in control and will respond in their usual ways if children and teens act out. You can let your child or teen know that it is normal to feel a lot of different emotions during this time (worried, sad, lonely, angry, bored), but that they still need to control their behavior.

Help your child stay physically fit. Find ways for your child and family to be physically active (go for a walk or bike ride if you are able to keep distance from others, find free family-friendly physical activity videos online). Teens may benefit from yoga, breathing, mindfulness, and guided meditation exercises (<https://www.headspace.com/covid-19>, <https://blog.calm.com/take-a-deep-breath>). Your child is more likely to eat healthy if there are healthy foods in the house. The body and mind are connected, and your child's physical health will affect how they handle stress and worry.

Helping Children Cope with Stress and Worry during COVID-19:

Information for Families of Children with Congenital Heart Disease (Contd.)

Help your child stay connected with friends and family members. Use apps on your phone or tablet to help your child stay connected. Teens will likely do this on their own but children may need help setting up virtual play dates or video chats with grandparents.

Have your child talk to a mental health professional. Your child may be able to speak to a mental health professional (psychologist, social worker, therapist) using online video meetings, without having to leave the house. This is called telehealth. If your child or teen is experiencing a lot of anxiety, depressed mood, or changes in behavior (for example, panic attacks, crying a lot, behavior problems, trouble sleeping or not feeling hungry like they used to), talking to a mental health professional may help. To find a mental health professional, contact your insurance company for a list of mental health clinics that take your insurance. Call the clinics to find out if they provide therapy using telehealth and have experience working with children and teens. You can also call your child's primary care doctor or your child's cardiologist or heart doctor's office for suggestions of where to call. Many hospitals have social workers and psychologists on staff who may be available for telehealth or may be able to help you locate someone who provides telehealth services.

The links below have information about helping children and teens handle COVID-19 related anxiety:

Center for Pediatric Traumatic Stress: <https://www.healthcaretoolbox.org/about-us/14-healthcare-toolbox/tools-and-resources/602-covid19-children-and-families.html>

National Child Traumatic Stress Network: <https://www.nctsn.org/resources/parent-caregiverguide-to-helping-families-cope-with-the-coronavirus-disease-2019>

SAMHSA: <https://store.samhsa.gov/product/Talking-With-Children-Tips-for-Caregivers-Parentsand-Teachers-During-Infectious-Disease-Outbreaks/PEP20-01-01-006>

National Association of School Psychologists: [https://www.nasponline.org/resources-andpublications/resources-and-podcasts/school-climate-safety-and-crisis/health-crisisresources/talking-to-children-about-covid-19-\(coronavirus\)-a-parent-resource](https://www.nasponline.org/resources-andpublications/resources-and-podcasts/school-climate-safety-and-crisis/health-crisisresources/talking-to-children-about-covid-19-(coronavirus)-a-parent-resource)

Information for families of children with developmental disabilities: <https://www.smores.com/udqm2-covid-19-preparedness>

Information for families of children with autism: <https://afirm.fpg.unc.edu/supporting-individualsautism-through-uncertain-times>

American Academy of Pediatrics Video on Talking with Children about COVID-19: <https://www.youtube.com/watch?v=FcYZWiF3PNc>

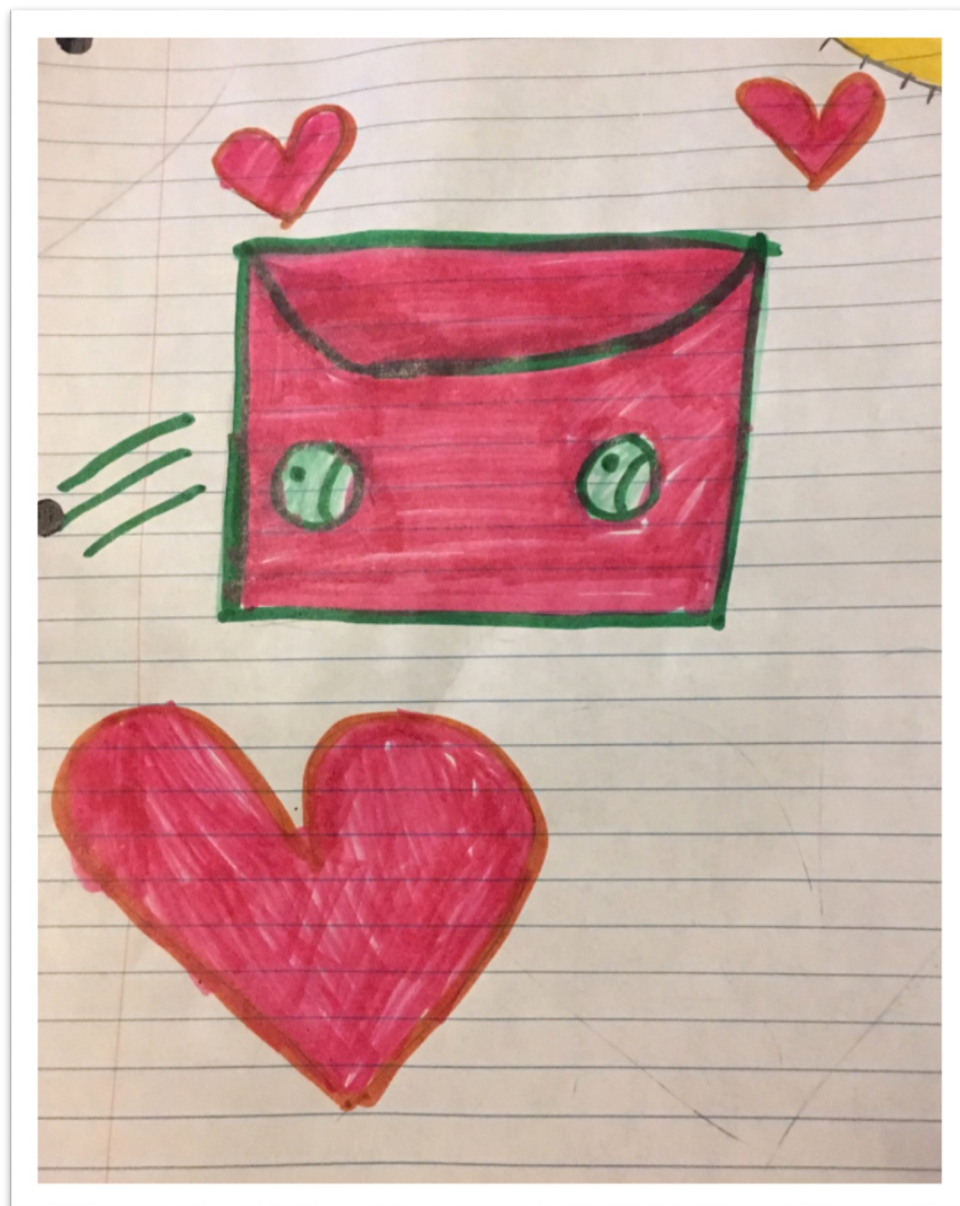
Online books/comics that help explain COVID-19 to young children: <https://akidsbookabout.com/>

<https://www.flipsnack.com/KeshetChicago/coronavirus-social-story/full-view.html>

<https://nursedottybooks.files.wordpress.com/2020/03/dave-the-dog-coronavirus-1-1.pdf>

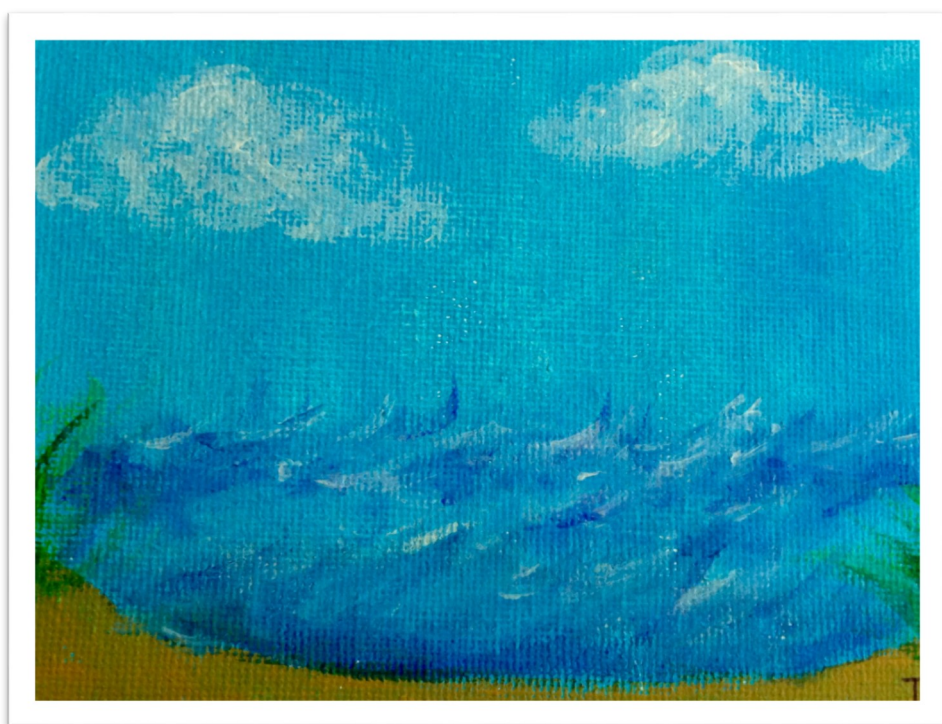
https://drive.google.com/file/d/1PYrKYfOBa4p-azI5z_46KJMbi1FSmL_Y/view

PHN ART GALLERY



-Analiz

PHN ART GALLERY



-Tess

FOR MORE INFORMATION...

If caregivers want to further discuss or receive other tips/guidance, please visit the blogs at the following sites:

Mayo Clinic: <https://connect.mayoclinic.org/>

The Clinical Trials.gov: <https://clinicaltrials.gov/ct2/home>

The Children Heart Foundation: <http://www.childrensheartfoundation.org/advocacy>

Patient Rising: <https://patientsrising.org/blog>

Kids Wish Network: <https://www.kidswishnetwork.org/2014/01/emilys-wish-is-to-meet-justin-timberlake/>

Mended Little Hearts: <http://www.mendedlittlehearts.org/>

Sisters by Heart: <http://www.sistersbyheart.org/content/blog>

To the essential staff on
the front lines

THANK YOU



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PARENTS....

Do you have a story to share about your experience in the SVR Extension Trial? Or a tip for other parents of children with HLHS/Single Ventricle?

Please spread the word! Send your SVR Extension Study Coordinator your story and/or ideas! Your feedback could be featured in a future issue of the SVR Extension Study Newsletter!