Heart History: Wonder Drugs: Prostaglandin E1- The elixir of life for babies with congenital heart defects (CHD)

Over the past century, there have been a number of “wonder drugs” whose discovery and development have truly changed the world. Think of ether, insulin, penicillin, to name a few. In pediatric cardiology and the world of babies born with certain complex congenital heart defects (CHD), no other drug discovery has had greater impact on prognosis than that of prostaglandin E.

Prostaglandin was initially discovered in 1935 by Ulf von Euler from seminal fluid and was so named because of it was thought to derive from the prostate gland and was a substance that resulted in uterine contraction. It took several decades and much research by independent investigators in Sweden (Sune Bergstrom and Bengt Samuelsson) and England (John Robert Vane) to elucidate of the true nature of prostaglandin(s) as many hormone-like substances present in nearly all body tissue with varying properties, including regulating the contraction and dilation of smooth muscle. For infants born with certain complex congenital heart defects, the smooth muscle of particular interest is in the ductus arteriosus (DA).

To understand the importance of the discovery of prostaglandin(s), we need to review the role of this vessel, the ductus arteriosus (DA) in infants with CHD. The DA is a vessel present in fetal life that connects the aorta and pulmonary artery serving to direct blood from the main pulmonary artery to the descending aorta. After birth the DA usually closes in the first few days of life through the action of oxygen and withdrawal of prostaglandins that we now know keeps the DA patent in utero. The DA, however, can be a lifeline for the infants with certain types of CHD serving to bring much needed blood either to the lungs in those born with critical obstructions to pulmonary blood flow such as pulmonary atresia or to the body in those born with critical obstructions on the left side of the heart, such as hypoplastic left heart syndrome and severe coarctations or interruptions of the aorta. These heart defects are the so called “ductal dependent” ones because infants born with these defects are dependent on continued patency of the DA for survival. Although surgical procedures were available in the pre-prostaglandin era to provide blood to the lungs such as the Blalock Tausig shunt discussed in previous newsletters, with ductal closure, many infants could not survive to surgery or were in such poor condition that post-operative recovery was compromised.

Following the initial discovery of prostaglandin, many investigators began research on its biological activities in various tissues. In 1973, several investigators, including Coccani and Olley in Toronto demonstrated the ability of prostaglandin E1 (PGE1) and E2 (PGE2) to dilate the DA of fetal lambs. A few case reports of its life saving ability to open the DA in critically ill neonates with CHD followed providing moti-

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Nurses’ Notes

Sports Clearance Reminders

♥ If your child plans on playing any level sport or activity, he or she may require annual clearance to participate from their pediatrician and/or pediatric cardiologist.

♥ In July 2014, New Jersey’s Governor Chris Christie signed the Scholastic Student-Athlete Act requiring stringent physical screenings, precautions and education regarding cardiac screening and action plans for athletes.

♥ There is a multi-step requirement for clearance, therefore planning for your child’s appointment is essential to ensure that forms are submitted in a timely manner. We all lead busy lives and the goal is not to delay your child’s participation.

♥ Providing sports clearance forms at your child’s next appointment is ideal. If your child is overdue for their follow-up appointment, it may take up to two weeks before paperwork is completed by our office.

♥ Guideline for scheduling your child’s sports clearance appointment:

- Fall sport- June appt.
- Winter sport- Aug appt.
- Spring sport- Jan appt.
- Sign up for MyChart (mychart.atlantichealth.org) for correspondence with your Cardiologist regarding non-emergent student-athlete questions.

Nurse Highlight

Staff Highlight Anjali Chelliah, MD
Pediatric Cardiologist

Anjali Chelliah, MD is a board-certified pediatric cardiologist. She received her undergraduate degree from Harvard University in 2000 and MD at the University of Pennsylvania in 2005. She continued moving up and down the East Coast as she pursued specialty and subspecialty training at Boston Children’s Hospital, Columbia University Medical Center, Children’s National Medical Center, and the National Institutes of Health. Most recently, she has served as a faculty member and pediatric cardiologist at Columbia University Medical Center/Morgan Stanley Children’s Hospital in New York City. She is excited to join the team at Goryeb, whose cardiologists she has known for many years and some of whom mentored her at Columbia.

Dr. Chelliah specializes in imaging the heart through fetal and pediatric echocardiography, cardiovascular MRI and CT and of cardiovascular 3D printing. She has published research articles on topics such as advancing the use of low-radiation cardiac CT in children with congenital heart disease (CHD), using 3D printers to create detailed models of patients’ hearts to guide their CHD surgery, and using fetal echocardiography to improve outcomes for babies with CHD. She also has a particular interest in global pediatric health. She has been fortunate to participate in several mission trips, including teen HIV prevention programs in India, a cardiac surgical mission in Uganda, and three months spent as a primary care pediatrician in the mountains of Lesotho.

Dr. Chelliah is a New Jersey native who is happy to be back in her home state. She enjoys spending time with her husband and two young sons, hiking, cooking, reading, and participating in her church community. She is an avid trivia lover who has competed (though sadly never won) on several game shows, first as a child on “Where in the World is Carmen Sandiego?” and more recently on “Jeopardy!” and “Who Wants to be a Millionaire?”

Attention College Students!

Sign up for Umergency, an all-in-one emergency, health, and safety App for college students and families to help navigate through any emergency.

The Umergency App was developed from real life experiences to instill peace of mind to students, parents, and campus administrators by keeping most vital medical resources at their fingertips. Features have a built-in medical consent form, digital version of student’s health insurance card, medical diagnosis, medications and safety contacts. Feel safe and prepared. Join Umergency! www.umergencyapp.com

Treasure Chest Fun!

Boy Scout or Girl Scout troop looking to do a good deed? Mitzvah project looking for ways to help others? New or duplicate birthday gifts without a home?

Donate unwrapped toys or small increment gift cards to our Treasure Chest.

A treat to bring home for our patients after their appointment to recognize their effort and cooperation.

Treasure Chest Ideas:
- Coloring books
- Markers
- Playdough
- Small Puzzles
- Board Games — travel size
- Small stuffed animals
- $5.00 Target, Walmart, Dunkin Donuts, Friendly’s gift cards

Contact Cathy Cerutti, LCSW 973-971-8689 for more information
Special Heart Camp for Children & Teens

It is not too early to think about summer camp! Children ages 7 to 17 can attend camp with the reassurance that a qualified medical team is there to address campers’ cardiac concerns and needs. New friendships form and campers can stay connected throughout the year with others who have been through similar experiences. Camp is a win/win as independent skills are developed and horizons are broadened. Camp is available to all families regardless of ability to pay. Contact camps directly for an application for your child or teenager.

Did you know?

- Drinking water is the best drink to satisfy thirst
- Pack a water bottle whenever you go out to school, work, games especially when it is hot or you’re sweating to make up for fluids lost
- Water transports nutrients and oxygen to the cells, removes wastes, protects joints and organs and regulates body temperature
- NOT drinking enough water causes dehydration and makes the heart pump faster
- NOT drinking enough water causes headaches, tiredness, irritability, poor concentration, fainting
- If you feel thirsty, you are probably starting to dehydrate
- Urine that is light yellow in color is best, anything darker means you need to drink more
- Avoid sugary beverages in favor of plain water
- Recommended daily intake of water for children: Ages 4-8 years 5 glasses Boys 9-13 6 glasses Girls 9-13 5-6 glasses
- What is your daily requirement of water?

Songs of Love

Create the Medicine of Music for a child who is facing a lifetime of medical, physical, or emotional challenges. Each Song of Love is created by talented, professional singer-song writers for anyone up to the age of 21. The songs can be created in any style of music and in any language.

You can hear examples of ‘Songs of Love” and complete the Love profile sheet by visiting their website at songsoflove.org or pick up a brochure in our office.

A great birthday, holiday, or ‘pick me up’ gift!

Family Connection

“EVERYTHING I HAVE DONE IS BECAUSE OF MY DESIGN,” says 8 year-old Gabriel, an extraordinary young boy, with a creative and inquisitive mind. Whether he is memorizing the periodic table or reciting Einstein’s theory of relativity, Gabriel never let his diagnosis of hypoplastic right heart and three open heart surgeries get in the way. What he loves instead is life! His infectious smile and gift for learning all and everything there is to know energizes him and makes him happy.

Enter his cardiologist, Dr. Suzanne Mone, who had a little secret! A friend of a friend works at Rovio Entertainment. She reached out and shared Gabriel’s journey and love for the game. Lo and behold, at Gabriel’s next cardiology appointment, a package from Espoo, Finland was waiting for him. Nothing compared to the look on Gabriel’s face as he opened the package filled with you guessed it, a fun-filled bag of ANGRY BIRDS gifts. Gabriel’s dad wants to thank Dr. Mone “for her straightforward approach, professionalism and youth”. Gabriel dreams of traveling to Finland to capture the ultimate Angry Birds experience. He even sent them a job application!

Espoo, Finland. Inspired by multi-colored birds who try to save their eggs from green colored pigs, their enemies.

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Thank you Dr. Mone for extending the doctor-patient relationship outside the exam room. Thank you Gabriel for sharing your Zest for Life!

The Edward J. Madden Open Hearts Camp
Ages 8 to 16
Great Barrington, MA
www.openheartcamp.org

Hope with Heart Camp
Ages 7 to 17
Harriman, NY
www.hopewithheart.org
YOU’RE INVITED TO OUR PEDIATRIC CARDIOLOGY PARENT DISCUSSION GROUP!

Welcome and share in an open and relaxed atmosphere amongst other parents who are journeying and living with CHD. Learn new resources, share experiences, practice relaxation techniques, discuss parenting and developmental themes.

FOR PRENATAL PARENTS TO TEENS:

Time: 7:30 pm to 9:00 pm
Place: 55 Madison Ave. 1st floor conference room
BEGINNING 2019 – we will meet on TUESDAY evenings
Winter Dates:
January 8
February 5
March 5

WELCOME TO OUR NEW STAFF!

Welcome to Amy Schmitt, BSN, RN, CPEN. She is excited to join the Pediatric Cardiology team. Amy joins us from the Pediatric Emergency Department at Morristown Medical Center and has been with Atlantic Health System for 11 years.

Jamie Ventrella, CMA joins our team with experience in working with pediatrics and adults as a medical assistant for 8 years. We welcome Jamie as an asset to our families and team.

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vation for a clinical trial. The pharmaceutical company Upjohn subsequently began a collaborative study in 1976 to test the efficacy of PGE1 in maintaining patency of the DA in infants with “ductal dependent” CHD. 56 centers and 492 patients over a 3 and ½ year period were involved in this study. The results were astounding. The impact of prostaglandin E1 on the early prognosis can be appreciated by the change in survival of infants born with pulmonary atresia which doubled during the study period. PGE1 resulted in dramatically altering the prognosis, survival and stabilization of infants prior to neonatal surgical intervention and optimizing outcomes for these fragile newborns.

The use of PGE1 in infants with cyanotic CHD is now standard of care and the downhill spiral witnessed in these infants in decades past, a distant memory. For their work on the elucidation of the effects of prostaglandins, Bergstrom, Samuelsson and Vane received the Nobel Prize in physiology/medicine in 1982. Through their pioneering work and that of subsequent researchers, infants with “ductal dependent” congenital heart defects received a new lease on life, a true “wonder drug.”

CHD AWARENESS WALK ALWAYS A FUN TIME

The sun was shining down at the 11th annual CHD Coalition fundraising walk and family fun day at Darlington Park on 09/30/18. A fun filled time was had by all! CHD families, children, volunteers, and medical team members participated in games, crafts, and dancing to the fabulous DJ’s music. It was pure fun. The best news is $160,000.00 was raised for CHD research. Please come out next year and support their mission. The CHD Coalition can be contacted at: info@chdcoalition.org or (973)850-6320.