



QUEEN'S PULSE

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THE QUEEN'S CENTER FOR ADULT CONGENITAL HEART DISEASE

A multidisciplinary team that provides comprehensive long-term and acute care for all forms of congenital heart disease (structural birth defects in the heart) as well as co-existing acquired heart disease.

The Queen's ACHD Team:
(From left to right)

Back row: Andras Bratincsak MD, Peter Tsai MD, Darren Lum MD

Middle row: Corey Lum DO, Shawna Mano'i RN, Jessica Ann Cordeiro PPA

Front row: Anne Kemble Luo MD, Lyndsey Sakuda APRN

**QUEEN'S HEART
INSTITUTE**



**THE QUEEN'S
HEALTH SYSTEMS**

ADULT CONGENITAL HEART DISEASE

The Adult Congenital Heart Disease specialty came to life in the second half of the 20th century, as more and more children with congenital heart disease survived into adulthood. Before congenital heart surgery was an option, children with congenital heart disease, especially those with moderate to severe disorders, did not survive. With the advancement of surgical techniques, availability of cardio-pulmonary bypass, and the discovery of surgical solutions and palliations, these kids were able to have a chance. Some of the heart defects could be repaired and some of them palliated to the extent that the child had a reasonable life. As the surgical techniques and post-surgical care improved, so did the survival and quality of life of these children. By the end of the 20th century, surgical interventions on minor and moderate heart conditions had excellent survival rates, and even severe heart conditions could be managed well. Overall, about 80% of children with heart conditions survived to adulthood. Initially, these adolescents and young adults were treated by the pediatric cardiologist, who were well versed in the intricate details of congenital heart lesions and the management of post-surgical complications or residual defects. As the population aged, certain complications, conditions and situations emerged, that were less familiar to a pediatric cardiologist. Atrial fibrillation, pregnancy related issues, coronary artery disease, and cardiac complications of adult onset conditions were more familiar to adult cardiologists. By the second decade of the 21st century, it was clear that the growing body of adult congenital heart disease patients needed a dedicated, specialty trained physician familiar with congenital lesions, coexisting pediatric conditions, as well as adult onset disorders,

late complications of congenital heart defects and residual lesions.

In 2015, the American Board of Internal Medicine established the requirements to achieve board certification in Adult Congenital Heart Disease, so the ACHD subspecialty was officially born. In the first year, only about 50 physicians were able to take the examination and get certified. Since then, the subspecialty continued to grow and currently there

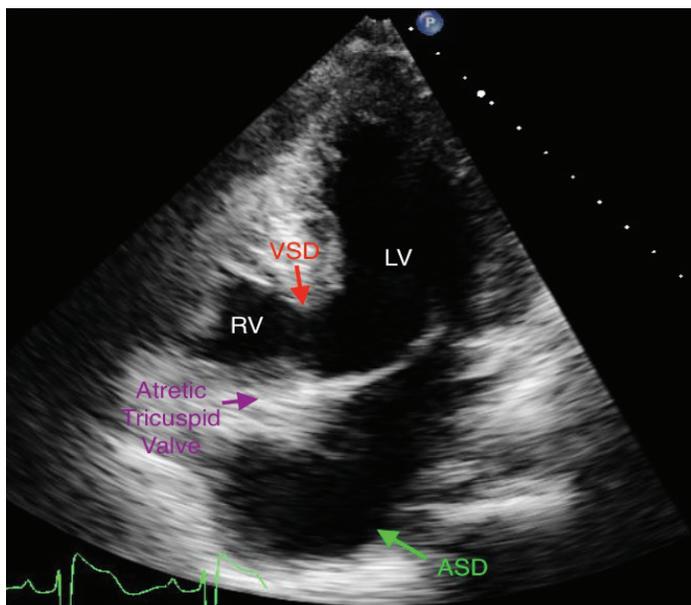
are a few hundreds of board certified ACHD specialists in the country and over 100 dedicated ACHD centers.

By 2020, more than 90% of children born with congenital heart disease will survive to adulthood. Congenital heart disease being the most common birth defect with a rate of about 1% of newborns, there are about 160-180 “new” ACHD patients graduating from childhood to adulthood every year in Hawaii. These patients require specialty care. Some of them have only one ventricle, similar to a fish heart, some of them had 5 open heart surgeries by the age of 18, some have an oxygen saturation of 80% at baseline, and many of them have significant restrictions, disabilities and comorbidities, truly requiring an entire center to take care of them.



Coarctation of the aorta, MRI: This patient presented at 28 years of age with hypertension and was found to have a bicuspid aortic valve. On exam, he had a 54 mmHg difference in systolic blood pressure between the right arm and leg. Interestingly, he had no bruit or thrill over the coarctation site and only a very soft continuous murmur in the intercostal spaces, due to minimal flow at the site of aortic coarctation and the presence of extensive collateral vessels. He was treated with stenting. *Yellow arrow: aortic coarctation. Red arrow: collateral vessels.*

In Hawaii, just as much as in other states with fruitful collaboration among physician groups and medical centers, ACHD care is a joint effort. Pediatric and adult cardiologists, electrophysiologist, interventional cardiologists, heart failure specialists, nurse practitioners, specialized sonographers, uniquely trained radiologists and a capable supportive staff ensures that these complicated patients receive appropriate care. Many of these patients require annual or biannual imaging studies, echocardiograms and cardiac MRIs, frequent cardiopulmonary stress tests and specialized cardiac catheterizations, not only to address emerging symptoms, but to provide comprehensive care to prevent complications, and with that keeping the ACHD patients as healthy as possible.



Tricuspid atresia (single ventricle), echo: This patient presented at age 23 with shortness of breath and an oxygen saturation of 59%. On exam she was cyanotic, had clubbing of the fingers and toes, and was in no distress. She was found to be anemic from menorrhagia, with a hemoglobin of 8 gm/dL. After correction of her anemia and treatment of menorrhagia, oxygen saturation rose to 77% and she reported no further symptoms. An expected hemoglobin in a patient with uncorrected single ventricle is closer to 18 gm/dL due to secondary erythrocytosis to compensate for chronic hypoxia. RV: hypoplastic right ventricle. LV: left ventricle. VSD: ventricular septal defect forming a functionally single ventricle. ASD: atrial septal defect forming a functionally common atrium.

As a team that integrates various subspecialties, hospital systems across the state, and also coordinates with ACHD centers on the mainland, the services the ACHD team provides include:

- Preventive and acute care in the outpatient and inpatient settings
- Specialized imaging (including echocardiography, CT and MRI)
- Electrophysiology consultation and procedures
- Specialized congenital cardiac surgery both locally and on the mainland
- Cardiac catheterization procedures and transcatheter (minimally invasive) therapies
- ACHD cardiac transplantation evaluation
- Support for primary care physicians caring for complex congenital patients
- Support for patients seeking to promote their cardiac health, including counseling on diet, exercise, family planning and prenatal care



Rhea Tiburcio PPA and Sara Hamele APRN

EXAMPLES OF CONDITIONS MANAGED:

- Atrial Septal Defect
- Ventricular Septal Defect
- Atrioventricular Septal Defect
- Bicuspid Aortic Valve
- Ebstein Anomaly
- Valve Atresia
- Tetralogy of Fallot
- Other Congenital Valvular Dysfunction
- Anomalous Pulmonary Venous Return
- Single Ventricle
- Eisenmenger Syndrome
- Fontan Circulation
- Transposition of the Great Arteries
- Truncus Arteriosus
- Coarctation of the Aorta
- Aortic Arch Anomalies
- Coronary Artery Anomalies



QUESTIONS?

Email our providers at
ACHD@queens.org



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LOOK
INSIDE
AND SEE
WHAT'S
NEW



MEET THE EXPERTS: THE QUEEN'S CENTER FOR ADULT CONGENITAL HEART DISEASE



Dr. Anne Kemble is a clinical cardiologist specializing in Adult Congenital Heart Disease and Echocardiography at the Queens Medical Center in Honolulu, Hawai'i. She trained in Adult Congenital Heart Disease with the Ahmanson/UCLA Adult Congenital Heart Disease Center. Dr. Kemble grew up in Honolulu, Hawaii, attended Brown Medical School in Rhode Island and completed training in Internal Medicine and Cardiology Fellowship at the University of Hawai'i, before joining the faculty at the University of Hawai'i Cardiovascular Disease Fellowship Program, where she has served as the Program Director and Associate Program Director. She is passionate about making high quality specialized care accessible to our local community.



Dr. Andras Bratincsak is a pediatric and adult congenital cardiologist and an associate professor of pediatrics at the John A. Burns School of Medicine in University of Hawaii. He is a native of Hungary, completed his medical school in Hungary followed by a PhD in neuroscience at the National Institutes of Health. After the pediatric residency at the University of Hawai'i he finished a pediatric cardiology fellowship program at the University of California in San Diego with subspecialty training in interventional cardiology, electrophysiology and adult congenital cardiology. He is board certified in pediatrics, pediatric cardiology and adult congenital heart disease. He has a special interest in heart rhythm problems and interventional cardiology.



Dr. Darren Lum is a diagnostic radiologist at the Queens Medical Center in Honolulu, Hawaii, specializing in Cardiovascular MR and CT. Dr. Lum was born and raised in Honolulu, Hawaii. He received his undergraduate education at Stanford University. He then attended medical school at the University of Hawaii John A. Burns School of Medicine. Dr. Lum completed his Radiology residency and a one-year fellowship in Magnetic Resonance Imaging at the University of Wisconsin-Madison. He is passionate about utilizing state of the art imaging technology to provide high quality cardiovascular imaging for the people of Hawaii.



Dr. David Singh specializes in cardiac electrophysiology. His clinical interests center on catheter ablation of supraventricular and complex arrhythmias. He earned his medical degree at Georgetown University School of Medicine, and completed Internal Medicine residency training at the University of California, San Francisco (UCSF), and trained in Cardiovascular Diseases at Cedars Sinai Medical Center in Los Angeles. In 2009, he returned to UCSF to undertake training in Electrophysiology. He also has helped to spearhead a number of international health initiatives, and maintains a passion for working with underserved populations. He has served as Chief of the Department of Cardiovascular Diseases for the Queens Medical Center since 2019.



Dr. Corey Lum is an invasive clinical cardiologist who specializes in Advanced Heart Failure & Transplant Cardiology at the Queen's Medical Center in Honolulu, Hawaii. Dr. Lum trained in Advanced Heart Failure & Transplant Cardiology at the University of California, Los Angeles (UCLA) Ronald Reagan Medical Center. He was born and raised in the Bay Area, California and moved to Hawaii in 2012. He is an Associate Professor of Clinical Medicine at the University of Hawaii, John A Burns School of Medicine and his research interests include non-invasive blood tests to evaluate for rejection in heart transplant recipients. Dr. Lum has a passion for heart transplantation, particularly in the adult congenital cardiac population given his training at UCLA and bringing heart transplantation back to Hawaii in the future.



Dr. Dip Banerjee is the Advanced Heart Failure director at Queens Medical Center, an Associate Professor of Medicine at the University of Hawai'i, and the Program Director of the University of Hawai'i Cardiovascular Disease Fellowship Program. He trained in Advanced Heart failure and obtained a Masters degree in clinical epidemiology at Stanford, where he was the medical director of the mechanical circulatory support program. At Queens he cares for patients with all forms of heart failure, and spends time in both the echocardiography and catheterization laboratories, making sure patients with end stage heart failure have all available options. He is the principal investigator for numerous multi-center clinical trials.



Dr. Peter Tsai is a cardiothoracic surgeon who trained at Baylor College of Medicine/Texas Heart Institute and specializes in surgical treatment of heart diseases involving the coronaries, valves and aorta, as well as general thoracic diseases. He is a Professor of Surgery, Division Chief of Cardiothoracic Surgery at the John A. Burns School of Medicine, and Medical Director of Cardiothoracic Surgery at Queen's Heart. He is passionate about education, research, and clinical outcomes. He advocates for an integrated cardiology-cardiac surgery service line to provide open, minimally invasive and endovascular treatment options for congenital, structural heart, coronary, arrhythmia, and aortic diseases.



Sara Hamele APRN has worked in electrophysiology since 2012 and in ACHD since 2017. She earned her Bachelor of Sciences in Exercise Science (kinesiology) from Western Washington University, and Masters of Science in Nursing from Pacific Lutheran University. Her clinical interest and expertise includes: adult congenital heart disease, cardiac device rhythm management, and arrhythmias; she also has experience in general cardiology and interventional cardiology. She enjoys the multidisciplinary nature of caring for ACHD patients, which incorporates her diverse cardiac training.



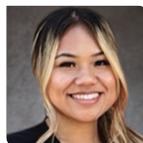
Lyndsey Sakuda APRN joined the ACHD team in 2020. She grew up on Oahu, where she completed a Master's of Science in Nursing at The University of Hawaii Manoa in 2012. She worked as a Registered Nurse in the Neonatal Intensive Care Unit at Kapiolani Center for Women and Children before pursuing her career in adult cardiology with the Queen's Heart Institute in 2015. Her clinical interests include adult congenital heart disease and general cardiology, patient advocacy, and ensuring patients with complex heart disease have access to the health care resources they need.



Shawna Mano'i RN is the Adult Congenital Heart Disease and General Cardiology Patient Care Coordinator for Queen's Heart Institute (QHI). She was born and raised on Oahu. She completed her Bachelor of Science in Nursing at Chaminade University of Honolulu, then joined the Queen's Medical Center's Comprehensive Cardiac Care Unit (CCCU) as a unit secretary, and worked as a telemetry RN through 2018. She also completed the Cardiac ICU program and has worked as an outpatient case manager for the cardiac service line, and in addition holds two associate degrees in Fashion Marketing and Apparel Manufacturing Management.



Jessica Ann Cordeiro is a physician practice assistant for the Queen's Heart Institute (QHI) general cardiology clinic and Adult Congenital Heart Disease program. She completed her Associates of Applied Science degree, and achieved her medical assistant and EKG technician certifications from Hawaii Medical College. She externed at the Queens Heart Institute, then soon after joined the team full-time with our general cardiology and ACHD staff. She enjoys spending time with her family, being creative, and going on adventures.



Rhea Tiburcio is a physician practice assistant for General Cardiology and the Adult Congenital Heart Disease program at Queens Heart Institute. She was born and raised on Oahu. In 2014 she began her liberal studies at UH West Oahu, then later found her passion in the medical field. She achieved her medical assistant certification from Hawaii Medical College. She completed her externship at Hawaii Vein Center, and soon after joined the QHI team. In her free time she enjoys going to the gym, traveling, and spending time with family and friends.