

Clinical Program Highlight

CONGENITAL DIAPHRAGMATIC HERNIA PROGRAM



Rebecca Stark, MD, FACS
Assistant Professor
Division of Pediatric General Surgery

Congenital diaphragmatic hernia (CDH) is a congenital anomaly characterized by failure of formation of part or all of the diaphragm. This allows abdominal viscera to herniate into the thorax, causing pulmonary hypoplasia and pulmonary hypertension. There is a wide spectrum of disease severity; in the most severe cases the mortality rate is as high as 80%. In that cohort, extracorporeal life support (ECLS) is commonly employed as a bridge to stabilize pulmonary hypertension and allow for lung development. Optimal outcomes for these babies necessitate a dedicated multidisciplinary care team, clinical expertise, long term follow up, and institutional experience. Institutions are considered high volume centers if they treat over 10 CDH patients per year. At Seattle Children's Hospital (SCH), we treat an average of 25 new patients per year with this condition, beginning in the prenatal period and extending throughout childhood. The average length of stay and resource utilization for these patients is high; they are some of the sickest patients in the hospital and benefit from standardization of care

whenever possible. To that end, Dr. Dan Ledbetter (late Professor of Surgery) developed a clinical guideline several years ago, which facilitated consistent ventilator management for these complex babies, and defined criteria for ECLS.

In 2019, we were able to build upon Dr. Ledbetter's foundation to start a comprehensive, multidisciplinary CDH program, led by Dr. [Rebecca Stark](#), Assistant Professor of Surgery and Dr. [Bob Digeronimo](#), Professor of Pediatrics, Division of Neonatology, with the goal of improving outcomes for these patients and their families. Our program spans all phases of pediatric care from the prenatal period through birth, through the neonatal intensive care unit (NICU) and remainder of the initial inpatient hospitalization, and in our long-term CDH clinic, which extends outpatient follow up through teenage years. Our multidisciplinary team is comprehensive, and includes pediatric general and thoracic surgery, neonatology, cardiology, pulmonology, nutrition, maternal fetal medicine, anesthesia, social work, genetic counseling,

respiratory therapy, and nursing. We have been able to significantly improve our outcomes by implementing innovative treatment modalities, and through consistent multidisciplinary collaboration and continuity of care.

Given the vast majority of CDHs are detected on screening prenatal ultrasound, we began by standardizing imaging for fetuses with this anomaly, including performing fetal MRI at 20 and 32 weeks gestation (Figure 1). We can then use the lung volume measurements on MRI to predict disease severity, need for ECLS, and survival, and to optimally plan the timing of delivery and urgency of transport from the delivery hospital to SCH. Moms whose fetuses have CDH receive comprehensive counseling from our multidisciplinary team at our [Fetal Care and Treatment Center](#), such that they can be as prepared as possible for their baby's clinical needs. On arrival to SCH, they are immediately cared for by our subspecialty CDH team of neonatologists and pediatric surgeons, including early institution of ECLS support within minutes of arrival for the most severe babies (Figure 2), who



Figure 1 Fetal MRI showing a severe left CDH



Figure 2 Chest X-ray showing left CDH with ECMO cannulas

Clinical Program Highlight

then undergo repair of their diaphragmatic defect within the first 24 hours on extracorporeal membrane oxygenation (ECMO). Further, we have developed novel protocols for use of bivalirudin in newborns on ECMO, early initiation of pulmonary hypertension treatment under the guidance of Dr. [Delphine Yung](#), Associate Professor of Pediatrics, Division of Pediatric Cardiology, and have achieved longer, safer ECLS runs as a result of our standardized pathways.

Once patients are out of the NICU and on the ward they undergo intensive feeding therapy and oxygen weaning based on standardized pathways developed by our multidisciplinary team. All patients with CDH are followed in our long-term multidisciplinary clinic which is run by Dr. Stark, Dr. [Bre Kinghorn](#), Assistant Professor of Pediatrics, Division of Pediatric Pulmonology, and [Carrie Foster](#), ARNP, Congenital Diaphragmatic Hernia Program, SCH.

All babies are enrolled in a national registry, the CDH Study Group (CDHSG), which includes over 80 institutions and allows for meaningful research evaluating ways to improve outcomes for these babies. Current experience tells us if these patients survive the neonatal period outcomes are excellent in terms of quality of life and overall morbidity is low. Overall survival is 70% nationally and 85% at SCH, and survival in the mild to moderate cohort is nearly 100%. The vast majority of deaths occur in the severe group of patients and specifically in those requiring ECLS, thus innovations in the care for these babies largely target this severe group. For example, recent work demonstrated that fetal tracheal occlusion (FETO) significantly improves outcomes for babies with severe CDH without unacceptable morbidity to mom or fetus; we anticipate bringing this technology to our burgeoning Fetal Intervention Program which is a joint venture between UW Medicine and SCH, also lead by Dr. Stark with Drs. [Martin Walker](#) and [Bettina Paek](#), Division of Maternal Fetal Medicine. Through this pioneering work and ongoing refinements of multidisciplinary care, we anticipate that we will be able to offer ongoing improvements in survival and quality of life to these critically ill newborns and their families.



save the dates

Annual Peter K. Buehler Visiting Professorship in Plastic Surgery Lecture

April 1st, 2022

GUEST LECTURER



Dr. William Y. Hoffman

University of California, San Francisco
Professor of Surgery, Chief, Division of Plastic
and Reconstructive Surgery,
Stephen J. Mathes, MD, Endowed Chair in
Plastic and Reconstructive Surgery

Please contact psurged@uw.edu for information

Division of Cardiothoracic Surgery Annual Visiting Professor

May 12th & 13th, 2022

GUEST LECTURER



Dr. Chris Caldarone

Professor and Chief, Division of Congenital
Heart Surgery Baylor College of Medicine
Program Director, Congenital Cardiac Surgery
Residency Baylor College of Medicine,
Chief, Congenital Heart Surgery: Texas Children's Hospital

Please contact lpayne3@uw.edu for information