

CardiovascularUpdate

Cardiology, Pediatric Cardiology and Cardiovascular Surgery News Vol. 19, No. 1, 2021

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Pediatric Cardiology at Mayo Clinic Celebrates 50 Years

Pediatric Cardiology at Mayo Clinic in Rochester, Minnesota, was formally founded by James W. DuShane, M.D., in November 1969. By then, pediatricians with an interest in cardiology had been working at the clinic for some time; Dr. DuShane had been part of the team supporting the first open-heart surgeries by Mayo Clinic cardiovascular surgeon John W. Kirklin, M.D., in 1955, and was joined soon thereafter by William H. Wiedman, M.D.

Fifty years later, in November 2019, current and emeritus Mayo Clinic Pediatric Cardiology staff and trainees and their families gathered in Rochester to celebrate this occasion over dinner in Balfour Hall at Mayo Foundation House.

Jonathan N. Johnson, M.D., current chair of Pediatric Cardiology, introduced the event and welcomed the attendees, many of whom had traveled from other countries to attend the event. He recognized the emeritus consultants from the division present at the dinner, including Douglas D. Mair, M.D., and Co-Burn J. Porter, M.D., and recognized deceased consultants.



From left, William D. Edwards, M.D., A. Jamil Tajik, M.D., and James B. Seward, M.D., share a conversation at the event.



Umberto Garofalo, M.D., is a pediatric cardiologist at Garofalo Health Care in Parma, Italy, an alumnus of the Mayo Clinic pediatric cardiology training program, and a former member of the board of directors, Mayo Clinic Alumni Association.

Joseph A. Dearani, M.D., then-chair of Cardiovascular Surgery, opened with a history of Cardiovascular Surgery at Mayo Clinic. Dr. Dearani described the first surgeries by Dr. Kirklin using the prototype mechanical pump-oxygenator originally developed by John H. Gibbon, M.D., and subsequently modified by Mayo Clinic staff. The awe-inspiring presentation showed how those first procedures took cardiovascular surgery into a new era and required perseverance, belief and the hard work of a multidisciplinary team.

Pediatric Cardiology Mayo Clinic in Rochester, Minnesota

Jonathan N. Johnson, M.D., Chair
Michael J. Ackerman, M.D., Ph.D.
Jason H. Anderson, M.D.
Allison K. Cabalka, M.D.
Bryan C. Cannon, M.D.
Frank Cetta Jr., M.D.
Benjamin (Ben) W. Eidem, M.D.
Donald J. Hagler, M.D.
Patrick W. O'Leary, M.D.
Timothy M. Olson, M.D.
M. Yasir Qureshi, M.B.B.S.
Nathaniel W. Taggart, M.D.
Charlotte S. Van Dorn, M.D.
Philip L. Wackel, M.D.
Trista A. Brown, P.A.-C., M.S.

Affiliated pediatric cardiologists

Christen R. Chamberland, M.D.
Justin M. Horner, M.D.
Susan G. MacLellan-Tobert, M.D.
Joseph T. Poterucha, D.O.
Alex J. Thompson, M.D.
Andrea G. Winters, P.A.-C.

Chairs of Pediatric Cardiology at Mayo Clinic in Rochester, Minnesota

James W. DuShane, M.D.
1969-1973

Donald G. Ritter, M.D.
1973-1984

David J. Driscoll, M.D.
1984-2004

Frank Cetta Jr., M.D.
2004-2017

Jonathan N. Johnson, M.D.
2017-present



Table display features photos of past consultants from Pediatric Cardiology.

Donald J. Hagler, M.D., consultant in Pediatric Cardiology, presented a history of the Cardiac Catheterization Laboratory, describing the earliest work in children. Dr. Hagler paid homage to former division chair Donald G. Ritter, M.D., for supporting and leading the Cardiac Catheterization Laboratory teams as they advanced the knowledge of cardiac disease in children.

A. Jamil Tajik, M.D., and James B. Seward, M.D., presented the history of the Echocardiography Laboratory from initial M-mode assessments to contemporary advanced imaging capabilities. The presentations by Drs. Hagler, Tajik and Seward showed how the laboratories worked together to characterize and publish numerous papers and texts on congenital heart disease. This synergy between the two laboratories continues today, and it has been a vital reason why Mayo Clinic remains at the cutting edge of invasive and noninvasive diagnostics and percutaneous therapies for patients with congenital heart disease. Drs. Tajik and Seward also recognized the critical collaboration with cardiac pathologist William D. Edwards, M.D.

Former chair Frank Cetta Jr., M.D., closed the evening with a reminder of what brings staff to Mayo Clinic: the patients. He thanked the many providers and staff who work tirelessly to care for Mayo's patients. He also recognized the efforts of David J. Driscoll, M.D., who served as the division chair for 20 years and trained many of the current staff. Dr. Driscoll's selfless leadership crafted much of the design of the current

division. The division extended particular thanks to those who traveled great distances to join the event, including alumnus Umberto Squarcia, M.D., of Parma, Italy. Dr. Squarcia trained at Mayo Clinic in the early 1970s, and in addition to maintaining close collaborative ties, he has served on the board of directors of the Mayo Clinic Alumni Association.

Discussion with attendees revealed that several important features of the division have remained consistent over the past 50 years. From 1969 to now, a constant has been the desire to give the best care possible to children every day.

The multidisciplinary nature of the program, bringing together colleagues from other specialties in the care of patients with complex congenital heart disease, has been and continues to be a formidable strength. There has always been a seamless sharing of data and knowledge among the anesthesiology, intensive care, pathology, pediatric and adult cardiology, and cardiovascular surgery teams. In particular, the strength of the cardiovascular surgery program has allowed the pediatric cardiology program to flourish, bringing new techniques and improved outcomes to patients.

Lastly, there is a constant connection between the present and the past. The division honors and thanks those whose hard work paved the way for others, and cardiology staff and trainees work every day to make them proud.

Pediatric Tracheal Surgery: A Multidisciplinary Team Approach to Management

Pediatric tracheal surgery is complex and requires an experienced multidisciplinary team that includes primarily specialists in pediatric cardiac surgery and pediatric otolaryngology, but also requires knowledgeable anesthesia, pulmonology and critical care staff. Nursing and respiratory therapy staff members also are essential. The team is typically involved with the preoperative evaluation, the operative approach and the postoperative management.

"Pediatric tracheal surgery may be required secondary to acquired tracheal disease, such as tracheal stenosis after tracheostomy or prolonged intubation, or congenital disease such as congenital tracheal rings, commonly associated with pulmonary artery sling," says Joseph A. Dearani, M.D., cardiovascular surgeon at Mayo Clinic in Rochester, Minnesota. Depending on the pathology, surgical management may include resection of the abnormal segment with an end-to-end anastomosis or a slide tracheoplasty. In general, when the pathology is less than 30% of the tracheal length (fewer than five tracheal rings), an end-to-end anastomosis is performed. Slide tracheoplasty can be used for more extensive pathology incorporating the majority of the trachea and even extending onto the bronchus.

Pulmonary artery sling is a rare vascular ring anomaly in which the left pulmonary artery arises from the right pulmonary artery and then passes around the distal trachea and right main bronchus coursing between the esophagus and trachea before exiting the pericardium to perfuse the lung. This "sling" causes anterior external compression of the esophagus and compression of the trachea. External compression of the central airways in severe cases may cause a ball-valve effect resulting in hyperinflation of the right lung. The majority of these patients (75%) have concomitant complete, circumferential tracheal rings as opposed to the normal membranous tissue of the posterior trachea, resulting in further narrowing of the airway.

"Tracheal surgery in children has evolved over the last three decades," says Joshua P. Wiedermann, M.D., pediatric otolaryngologist at Mayo Clinic in Rochester, Minnesota. "It was not until the 1960s that tracheal stenosis was classified and the first successful resection was reported."

Starting in the 1960s, Dr. Hermes Grillo led pioneering work at Boston Children's Hospital examining tracheal anatomy and developing techniques to improve outcomes of tracheal

resection. Many of the advances in this field began with the first successful pulmonary artery sling repair in 1953, followed by pericardial patch tracheoplasty in 1982, and then the "autograft" technique in which a piece of the patient's own trachea was used for the reconstruction. In more recent years, the slide tracheoplasty has become the established technique for long-segment tracheal pathology.

Slide tracheoplasty is performed via a median sternotomy with the use of cardiopulmonary bypass, which provides ventilation and oxygenation without the need for cross-table ventilation. Any intracardiac lesions are repaired while on cardiopulmonary bypass prior to opening the trachea. Repeat bronchoscopy is performed to approximate the length of the tracheal pathology. The trachea is dissected free with attention to preservation of its blood supply posterolaterally. Once the trachea has been mobilized, a needle is inserted at various points through the trachea with simultaneous bronchoscopy to confirm the superior and inferior extent of resection required. The trachea is then sharply entered at the midpoint of the stenotic segment, and the trachea is transected. In the case of a slide tracheoplasty, two counter incisions are made with the one on the superior portion of the trachea performed anteriorly and the inferior portion of the trachea performed posteriorly (Figure 1, page 4). Each incision continues until normal trachea is encountered. The corners of these incisions are rounded and the two portions are slid together such that the trachea becomes half as long but with a twofold increase in diameter. The tracheal segments are then sutured together with attention to create a tension-free repair.

The resultant tracheal anatomy is examined intraoperatively via bronchoscopy to ensure a satisfactory result, the airways are suctioned, and the patient is reintubated. The repair is tested by filling the mediastinum with saline and providing progressively increased airway pressure to a maximum of 30 mm Hg. Once the repair has been confirmed to be satisfactory, the patient is weaned from cardiopulmonary bypass.

In cases with pathology less than 30% of the tracheal length (fewer than five tracheal rings), the trachea is mobilized and the extent of the pathology is similarly localized using bronchoscopy. The abnormal tracheal rings are resected and end-to-end anastomosis is performed (Figure 2, page 4).



Joseph A. Dearani, M.D.



Elizabeth H. Stephens, M.D., Ph.D.



Joshua P. Wiedermann, M.D.

Cardiovascular Surgery Mayo Clinic in Rochester, Minnesota

Juan A. Crestanello, M.D., Chair
Arman Arghami, M.D.
Gabor Bagameri, M.D.
Richard C. Daly, M.D.
Joseph A. Dearani, M.D.
Kevin L. Greason, M.D.
Vishal Khullar, M.B.B.S.
Prasad Krishnan, M.D.
Alberto Pochettino, M.D.
Phillip G. Rowse, M.D.
Hartzell V. Schaff, M.D.
Elizabeth H. Stephens, M.D., Ph.D.
John M. Stulak, M.D.

Pediatric Otolaryngology Specialty Group Mayo Clinic in Rochester, Minnesota

Shelagh A. Cofer, M.D., Chair
Laura J. Orvidas, M.D.
Joshua P. Wiedermann, M.D.

The resultant tracheal repair is examined with bronchoscopy and the patient is similarly suctioned and reintubated, the anastomosis is tested, and the patient is weaned from cardiopulmonary bypass. Short, isolated cervical segments often do not require sternotomy or cardiopulmonary bypass.

Pulmonary artery sling repair with concomitant complete tracheal rings uses the tracheal surgical techniques detailed above; however, the left pulmonary artery reimplantation into the distal main pulmonary artery is performed prior to the tracheal repair. Care is taken to trim the length of the left pulmonary artery to ensure it does not kink in its course to the hilum.

Priorities during the postoperative management include maintaining a tension-free repair, adequate pulmonary hygiene, and prevention and management of granulation tissue at the anastomosis. While some groups have routinely used a “chin” or “Grillo” stitch to prevent neck extension and resultant tension on the anastomosis, this is not felt to be necessary by most groups involved with tracheal surgery.

“Results of tracheal surgery in experienced centers with a multidisciplinary team have been excellent and continue to improve for a very challenging population,” says Elizabeth H. Stephens, M.D., Ph.D., cardiovascular surgeon at Mayo Clinic in Rochester, Minnesota. In a study published in *Annals of Thoracic Surgery* in 2020 by Dr. Stephens, of 41 patients with a median age 4.1 months, including 22% who required intracardiac repairs and 20% who had lung agenesis or severe hypoplasia, there were three patients with an early mortality (one from respiratory failure, one with liver failure secondary to biliary atresia, and one secondary to aortic valve endocarditis). There was one late death secondary to sepsis. Interestingly, those with lung hypoplasia or agenesis did not experience worse outcomes, but these patients were more symptomatic prior to surgery, had a longer length of stay and longer duration of mechanical ventilation. Midterm follow-up was available on 26 of the 38 survivors, with a mean of 4.6 plus or minus 5.8 years. There were no re-operations in this follow-up period.

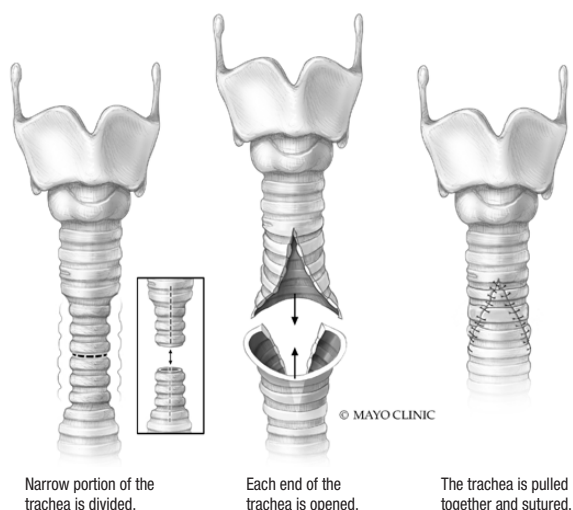


Figure 1. Slide tracheoplasty.

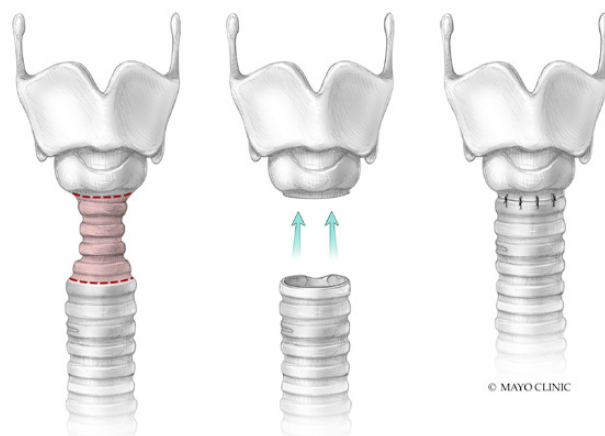
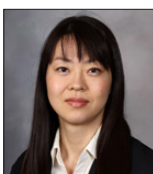


Figure 2. Tracheal resection with end-to-end anastomosis.

Honors



Nahoko Kato, M.D., Ph.D.

Nahoko Kato, M.D., Ph.D., advanced echocardiography research fellow from Tokyo Bay Urayasu Ichikawa Medical Center, Chiba, Japan, was selected as one of four finalists in the Arthur E. Weyman Young Investigator's Award competition of the American Society of Echocardiography (ASE). Her research project, “Projected Transmitral Gradient: A Novel Tool for Assessment of Mitral Stenosis,” was presented at the ASE 2020: Virtual Experience in August.

In conclusion, pediatric tracheal surgery is a challenging field requiring the close collaboration between the pediatric cardiac surgeons, a pediatric otolaryngology airway specialist and a multidisciplinary team. In tertiary care centers with comprehensive expertise, the current techniques of simultaneous tracheal and cardiac repairs have yielded excellent early and late results.

For more information

Backer CL, et al. A history of pediatric tracheal surgery. *World Journal of Pediatric Congenital Heart Surgery*. 2010;1:344.

Stephens EH, et al. Pediatric tracheal surgery: A 25-year review of slide tracheoplasty and tracheal resection. *Annals of Thoracic Surgery*. 2020;109:148.

Transforming Cardiovascular Treatment for the Transgender Population

An estimated 1.4 million people in the U.S. identify as transgender, constituting 0.6% of the population. However, these numbers are likely conservative due to the broad range of terms used to describe transgender identities, and most notably, lack of data collection in population-based studies. Many surveys have been undertaken looking at the delivery of health care for people who identify as transgender and gender diverse. The numbers are sobering:

- 19% are uninsured.
- 19% have been refused care due to their gender-nonconforming status.
- 28% have postponed necessary medical care due to discrimination by medical providers.
- 33% delayed or did not try to get preventive health care due to discrimination by medical providers.
- 50% reported teaching their medical providers about transgender care.

The lack of knowledge has been identified as the largest barrier to health care for transgender individuals.

The percentage of individuals who identify as transgender among adults is highest in those ages 18 to 24, at 700 per 100,000 (0.7%), compared with 600 per 100,000 (0.6%) in those ages 25 to 64, and 500 per 100,000 (0.5%) in those age 65 and older. According to U.S. Transgender Survey

results, by age 20, 94% of respondents began to feel that their gender was different from their sex assigned at birth, 73% of respondents began to think that they were transgender, and 52% began to tell others that they were transgender. Transgender men are assigned female sex at birth, while transgender women are assigned male sex at birth. The proportion of transgender women to transgender men is reported as high as 2-to-1. However, these ratios should not be taken as a definitive indication of population sizes, given the limitations in methodology used to record them.

Transgender patients have higher rates of health concerns that negatively impact their cardiovascular (CV) health, including depression, substance abuse, tobacco use, obesity and lack of health care. In regard to cardiovascular disease (CVD) mortality and morbidity in the transgender population, studies have indicated that the mortality rate from CV causes in transgender females is higher than in cisgender females (1.64 to 2.11 with a 95% CI, 1.32 to 3.21), although similar to that in cisgender males. The mortality rate has improved as oral estrogen dosing has decreased or been switched to transdermal formulations. The mortality rate in transgender males is similar to that of cisgender females. In small European studies, transgender women seem to have a higher CVD risk than do transgender males. Transgender males using testosterone do not seem to have



Rekha Mankad, M.D.



Kari A. Dessner, A.P.R.N., C.N.P.

Honors



Abhishek J. Deshmukh, M.B.B.S.

Abhishek J. Deshmukh, M.B.B.S., and Guy S. Reeder, M.D., cardiologists at Mayo Clinic in Rochester, Minnesota, were selected as 2020 Teachers of the Year by the fellows in Cardiovascular Medicine. This award is presented to those educators who have demonstrated a superior and compassionate commitment to education, mentoring and patient care.



Guy S. Reeder, M.D.

an increased CVD risk, which would not follow the typical tendencies of testosterone use in cisgender males.

There is a paucity of large, randomized trials to assess the CVD risk in transgender women. Common limitations of the studies available include small cohorts, study design and study length. In one electronic medical record-based cohort study, transgender female participants had a higher incidence of venous thromboembolism (VTE), with two- and eight-year risk differences of 4.1 (95% CI, 1.6 to 6.7) and 16.7 (CI, 6.4 to 27.5) per 1,000 people relative to cisgender men and 3.4 (95% CI, 1.1 to 5.6) and 13.7 (95% CI, 4.1 to 22.7) relative to cisgender women. The overall analyses for ischemic stroke and myocardial infarction demonstrated similar incidence across groups. More pronounced differences for VTE and ischemic stroke were observed among transgender female participants who initiated hormone therapy during follow-up. Other smaller studies have had similar results.

Mayo Clinic developed a multidisciplinary Transgender and Intersex Specialty Care Clinic in Rochester, Minnesota, in 2015. The mission of the clinic is to provide specialty, multidisciplinary, person-affirming care for transgender, gender-diverse and intersex individuals in a safe and respectful environment while optimizing physical, emotional and social health. The vision is to become a premier center for transgender health, serving transgender, gender-diverse and intersex individuals by:

- Providing the Mayo Clinic Model of Care to our patients
- Sharing knowledge with providers, learners, patients and the community to promote better health for patients
- Advancing the science of transgender medicine through scholarly activity

The Women's Heart Clinic has partnered with the medical and surgical teams of the Transgender and Intersex Specialty Care Clinic to provide culturally sensitive cardiovascular risk assessments, as well as assessment of the cardiovascular risk of gender-affirming surgery. The Women's Heart Clinic uses the most up-to-date information available to

best meet the needs of the patients. A multifaceted approach using questionnaires, risk assessment tools and laboratory data is used in conjunction with the comprehensive review by the Transgender and Intersex Specialty Care Clinic social worker to best look at the seven determinants of health and how they apply to the cardiovascular health of each patient. Electrocardiography, stress testing and echocardiography are used to further assist with the overall assessment in those individuals at CV high risk as well as those with symptoms suggestive of CVD. Dietitians and exercise specialists are available to assist with specific questions. However, it is important to recognize that normative data for the transgender population is lacking. This limited information underscores the need to collect as much data as possible to allow for comprehensive discussion and evaluation.

Gender-affirming surgeries can be part of the health care experience for transgender patients. Reviews of outcomes have indicated that, in the past, CV events were seen more commonly in patients undergoing gender-affirming genital surgery than in the cisgender general population undergoing surgical procedures of similar risk. Major perioperative concerns facing transgender patients include VTE such as deep vein thrombosis and pulmonary embolism; the rate is reported as high as 6%. Based on the most recent evidence from a high-volume center, holding gender-affirming hormone therapy for two to four weeks pre- and postoperatively mitigates 50% to 70% of the rate of VTE. Additionally, utilizing comprehensive CV risk assessment tools and cardiac care preoperatively mitigates much of the rest of the increased rate of events.

The appropriateness of hormone replacement therapy (HRT) in the general population is one of the indications for consultation to the Women's Heart Clinic. Given the adverse findings of the Women's Health Initiative (WHI), there is greater need for discussion about implementation of hormone therapy for menopausal women. One of the key findings from the WHI is that cisgender women over age 60 have a greater risk of CV events when initiated on HRT, thus the notion

Honors



Regis I. Fernandes, M.D.

Regis I. Fernandes, M.D., cardiologist at Mayo Clinic in Phoenix/Scottsdale, Arizona, has been selected for the Fellow of the National Lipid Association (NLA) award. This award is reserved for NLA members who have made significant regional and national contributions to the science and practice of clinical lipidology and to the organizational mission.

that “aged” vasculature has an adverse response to exogenous estrogen therapy. As cisgender males have greater incidence of atherosclerosis than do cisgender females of the same age, the risk of feminizing hormones in older transgender women must be recognized and appropriately addressed. The Women’s Heart Clinic has expertise in the discussion about the benefits and risks of HRT for cisgender women. Thus, it is logical to be the place where transgender women can be evaluated for their own appropriateness of initiation of gender-affirming hormone therapy, as well as preoperative risk and longitudinal risk assessment across all age groups.

The priorities for research on barriers to transgender health care must include determination of the gaps in knowledge among the provider workforce across the range of training, potential interventions for those gaps, determination of indirect barriers such as environment and stigma, and potential solutions to overcome those barriers. Several papers recommend looking at the length of time a patient has been on hormone therapy to determine whether to use natal sex or affirmed gender to determine cardiovascular risk, medication therapy and imaging parameters. However, specific guidelines do not exist. We are currently collecting data to help determine guidelines for imaging.

Our cardiac rehabilitation specialists have completed a survey of the transgender patients that they have seen so they can best provide culturally sensitive care. The patients were forthcoming with the positive and negative experiences they have encountered. Based on this information, changes have been made to the physical cardiac rehabilitation area as well as to the instruction provided. The changes have been well received by the transgender patients as well as their cisgender counterparts.

The National Academy of Medicine has identified transgender adults as an understudied population in critical need of health research. In the U.S., there has been little attempt to determine the specific workforce needs to provide care or to determine the current status of that care. The interest is now being recognized, but it is outstripping the available science. Research

needs to be done carefully so that suboptimal methods do not beget suboptimal science.

For over 20 years, the Women’s Heart Clinic at Mayo Clinic’s Rochester campus has been a national and international leader in the provision of evidenced-based, gender-specific care to women with heart disease. The clinic was formed because of an identified disparity in the quality of cardiac care available for women. We have a strong history of sex-based clinical research, knowledge of hormone therapy and cardiovascular risk, and dedication to providing care to an underserved population. The providers of the Women’s Heart Clinic are uniquely qualified to advance the cardiovascular care of transgender patients and are committed to using their knowledge and resources to do so.

For more information

2015 U.S. Transgender Survey. National Center for Transgender Equality. <http://www.ustranssurvey.org>.

Women’s Heart Clinic Preventive Cardiology Mayo Clinic in Rochester, Minnesota

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Aida N. Lteif, M.D.

Jorys Martinez-Jorge, M.D.

Diana M. Orbelo, Ph.D.

Justine S. Herndon, P.A. -C.

Honors



Samuel J. Asirvatham, M.D., electrophysiologist at Mayo Clinic in Rochester, Minnesota, was awarded the 2020 Mayo Clinic Distinguished Inventor Award. This award is presented annually to a member of the Mayo Clinic voting staff whose career demonstrates evidence of great distinction in innovative and impactful contributions to the betterment of human health through successful invention of novel health care solutions.

Samuel J. Asirvatham, M.D.

Mayo Clinic

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