January 2nd, 2019

Dear Colleague,

The Inova Advanced Lung Disease and Transplant Program has enjoyed another successful year with 26 lung transplants in 2018. Based on the most recent data from The Scientific Registry of Transplant Recipients (SRTR), our average wait time for a transplant is 3.7 months, while our survival statistics continue to be in line with the National averages; one month survival of 98.11% (U.S 97.15%), one year survival 87.91% (U.S. 88.02%) and three survival 66.37% (U.S.69.77%).

A new lung allocation system came into effect November 2017 which offers lungs out in a 250 mile radius based on the site of the donor hospital. This system has placed us in direct competition with other large programs in the East for available donor lungs. Fortunately, this has not affected us adversely, but has rather been a "mixed blessing". On the plus side our numbers have increased slightly, but the downside is that generally our patients' diseases have to advance and they have to get sick enough to score high enough to draw lung offers. The net result is that the patients we do transplant are generally quite a bit sicker and certainly in greater need for a transplant. For example in 2017, 4 of 24 patients were waiting in-house for donor lungs, while this past year 21 of 26 recipients were in-house at the time of their transplants, reflecting the overall greater severity of disease illness. In addition 4 of our 24 patients this year were transplanted off ECMO support versus 1 patient in 2017. Additional downsides are that we have to travel further to retrieve lungs, with all but 2 of the donor lungs this year coming from outside of our local region. Sicker patients and longer ischemic times are invariably associated with increased primary graft dysfunction and greater hospital lengths of stay.

There are also certain diseases (notably COPD), where it is difficult to generate a high score, no matter how compromised. Invariably lungs that do get offered to these patients are "marginal" and have been turned down by others. To meet the demand for donor lungs, we now have ex-vivo lung perfusion available to improve marginal lungs. This is being orchestrated through a research protocol in collaboration with Lung Biotechnology, a subsidiary of United Therapeutics (Silver Spring, MD). We have had 2 EVLP runs this year; one lung was successfully transplanted, while the other lung didn't pan out. This latter "run" was also done in conjunction with our first "DCD" (donation after cardiac death) procurement.

Our Advanced Lung Disease Program continues to grow with a record number of new referrals this past year (n=698), and 493 new evaluations, which is a 28% increase over our prior record year in 2012. On November 14th of this year we were recognized as a World Association for Sarcoidosis and other Granulomatous Diseases (WASOG) Clinic. We are now one of 75 CMS accredited lung transplant centers, one of 52 accredited Comprehensive Care Centers for Pulmonary Hypertension, one of 60 Pulmonary Fibrosis Foundation Care Centers, and one of 112 Cystic Fibrosis Foundation-accredited Care Centers in the United States. Throw in the WASOG accreditation and we are now only one of 18 to hold all of these designations.

We continue to welcome and encourage the referral of any patients with interstitial lung disease, pulmonary hypertension, cystic fibrosis, non-CF bronchiectasis, sarcoidosis, A1AT deficiency, rare lung diseases, not so rare lung diseases such as advanced COPD and any diagnostic dilemmas. Interestingly, about 14% of our new evaluations traveled >100 miles to see us and ~6.4% were from more than 200 miles away, reflecting our standing as a regional and national referral center. Between all our programs, we follow about 1526 patients (8% increase c/w 2017) with 3647 outpatient visits for the year (4.74% increase). We couldn't possibly see all these patients without the ongoing support of their referring providers and we therefore encourage co-management and care from patients' community Pulmonologists and other physicians.

Due to our ongoing growth, we were very happy to be able to justify and create a position for our former Advanced Lung Disease Fellow, Dr.Kareem Ahmad, who has stayed on staff with us. We are therefore up to 6 fulltime physicians, who have been with the Program a total of 52 years (SN-22; OS-13; AWB-8; CK 5, SA-2, KA-2). Our surgical team remains robust with Dr. Linda Bogar as our surgical director, and Drs. Liam Ryan, Eric Sarin and Ramesh Singh as our skilled transplant surgical team. Our long-

standing Social Worker Jane Harrison retired this year after about 30 years of service. Jane will be sorely missed but we have ongoing great SW support with Elizabeth Davies-Wellborn and Susan Perry, who recently joined us.

On the education front, we had 38 residents and fellows rotate with us during 2018. This year marked the third year of our Advanced Lung Disease and Transplant fellowship, with Dr. Matt Koslow joining us from July 2018 through June 2019. We were privileged to have a Pulmonologist from Brazil join us for a one month preceptorship and next academic year we will be welcoming a French Pulmonologist for a year of research with our ILD program. We also welcomed a one week observership for a Nurse Practitioner in the region. Our educational efforts have therefore become broader and more International with a number of case conferences orchestrated during the year with Pulmonologists in Brazil and South Africa. Our summer student research program has also continued to mature with multiple applications for our 5 available slots.

We hold monthly ILD multidisciplinary meetings as well as monthly pulmonary hypertension multidisciplinary meetings. These are held from 7:30-8:30 am EST on the 2nd Wednesday and 2nd Thursdays of the month. We invite all referring physicians to participate, even remotely via GoToMeeting, if you have any "tough" cases to share or if you just wish to solicit input from our group. If interested, please contact us and we can share the meeting invitation/link with you.

Other new programs to look forward to in 2019 are the inception of a CTEPH (chronic thromboembolic PH) program under the directorship of Drs. Oksana Shlobin and Linda Bogar. Both have been out to San Diego to undergo training already with other team members (including Dr. Chris King-Associate Medical Director and Dr. Melanie Atkins- Thoracic Radiology). This program will include a balloon angioplasty component with Dr. David Spinosa (IR) and Dr. Benham Tehrani (Cardiology) due to undergo training in Japan in 2019. We will also be initiating an endobronchial lung volume reduction program under the directorship of our Interventional Pulmonologist, Dr. Amit "Bobby" Mahajan. Endobronchial valves were approved a few months ago by the FDA and may be a viable option for select COPD patients. Consideration of this option is of increased importance given the reduced likelihood for COPD patients to receive donor organs with the new allocation system. In 2019 we will also be instituting pharmacogenomic testing (Medimap® panel-25 genes, 146 medications) as part of our clinical assessment to help anticipate any medication issues, limit adverse events and optimize clinical outcomes.

We have also continued to be very active on the research and writing front with 16 original research manuscripts (accepted or published),9 reviews, 1 editorial, 2 consensus papers, 2 case reports, and 34 abstract presentations at International meetings including the ATS, ISHLT, ERS and Chest 2018 meetings. Between us, we also delivered ≈15 talks at these same and other International meetings.

Please feel free to directly call or email any of our Pulmonologists with new referrals, questions, issues, or updates on existing patients (contact info next page). If any patients need to be seen expeditiously, then please call or email one of us directly and we will accommodate them earlier. We wish you, your staff and families a happy, healthy New Year. Thank you for your ongoing support and confidence in our Program.

With best wishes,

Steven D. Nathan

Steven Nathan, MD
Medical Director

INOVA ADVANCED LUNG DISEASE AND TRANSPLANT REFERRALS

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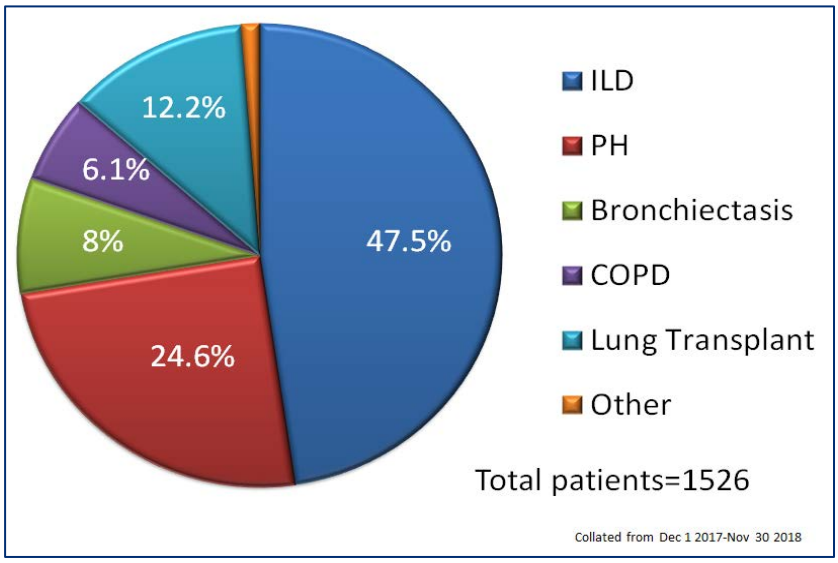
Inova Fairfax Advanced Lung Disease & Lung Transplant Program:

CLINIC COHORTS 2018



Oksana, Whitney, Shambhu, Steve, Chris, Kareem & Nargues

CONTACT INFORMATION

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Interstitial Lung Disease Program (by Chris King, MD)

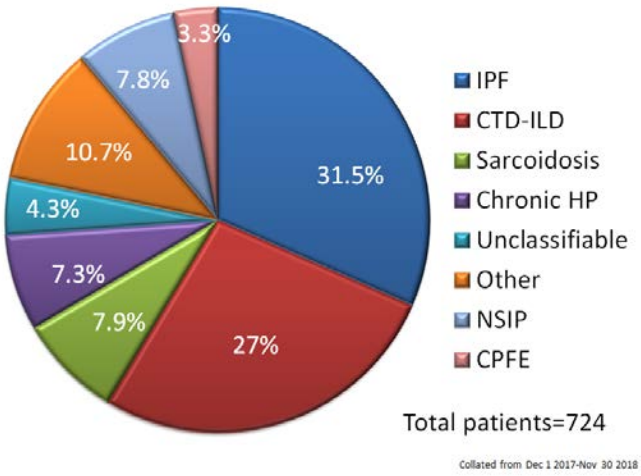
The past year saw many exciting developments in the interstitial lung disease clinic at the Inova Advanced Lung Disease (ALD) and Transplant Clinic. Our clinic finished as the second highest enrolling center in the United States for the Pulmonary Fibrosis Foundation Registry. It is hoped that this registry will provide vital information on the natural history of various interstitial lung diseases (ILD), specifically idiopathic pulmonary fibrosis (IPF). Dr. King and Dr. Nathan have already tapped into this wealth of data, performing an analysis of anticoagulant use in ILD and its impact on mortality. They are hoping to publish their findings in 2019.

Under the leadership of Dr. Shambhu Aryal, the Inova ALD Clinic has become a World Association of Sarcoidosis and Other Granulomatous Disease (WASOG) Accredited Sarcoidosis Clinic. While we feel our clinic has traditionally provided exceptional care for sarcoid parenchymal lung disease and sarcoid-associated pulmonary hypertension, we hope to expand our ability to provide comprehensive multi-organ system management of sarcoidosis in partnership with our colleagues in the Inova Advanced Heart Failure Program, as well as Dermatology, Neurology, and Otolaryngology.

Clinical research of IPF has remained a prominent focus of the ALD Clinic. Inova was a top enroller in a clinical trial of recombinant human pentraxin 2 in IPF which demonstrated a slower rate of decline in patients treated with the study drug in comparison to placebo with or without background antifibrotic therapy. Our own Dr. Whitney Brown was included as an author in the JAMA publication resulting from this study. (JAMA 2018; 319(22): 2299-2307) A number of promising trials for IPF have started enrolling or are slated to start in early 2019 including:
- SPIRIT (https://clinicaltrials.gov/ct2/show/NCT03573505?recrs=a&lead=biogen&rank=4) a weekly subcutaneous injectable therapy thought to reduce the rate of decline in forced viral capacity
- CLEANUP-IPF https://clinicaltrials.gov/ct2/show/NCT02759120 An NIH-sponsored study to assess the efficacy of either trimethoprim-sulfa or doxycycline on time to the first non-elective respiratory hospitalization or death. This pragmatic trial has very inclusive enrollment criteria and can be offered to nearly all patients with IPF.
- ISABELA https://clinicaltrials.gov/ct2/show/NCT03733444?recrs=ad&type=Intr&cond=ipf&phase=2&rank=3 Another phase 3 trial of an anti-fibrotic agent, a once daily tablet, in IPF sponsored by Galapagos NV.

As always please contact us if you have interest in referring patients for ILD care or consideration of enrollment in one of our clinical trials.

The Spectrum of ILD seen in the Inova ILD Clinic in 2018



Inova Cystic Fibrosis & Bronchiectasis Program (by A. Whitney Brown, MD)

We follow ≈56 patients with **non-CF bronchiectasis** from a range of causes such as immunoglobulin deficiency, prior infection, primary ciliary dyskinesia, and idiopathic. We use a multidisciplinary approach to care very similar to the care model in CF, and share many of the same resources and treatment strategies to ensure comprehensive care and best outcomes. In October, we were fortunate to hire a 0.5 FTE dietician, Erin Lopynski, to treat both our CF and non-CF bronchiectasis patients as we recognize the importance of nutritional status in these diseases.

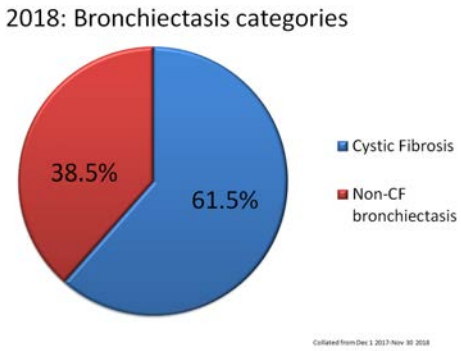
Our **Adult CF Program** continues to grow and flourish since our accreditation as a CF Foundation Care Center in January 2017. We began 2018 with approximately 65 CF patients and have assumed care for an additional 25 CF patients since that time (~38% growth!!).

With more patients comes… more opportunities. Given our increasing CF patient population, we are taking advantage of as many programs and grants from the CF Foundation as we can as:

- We are participating in a Learning and Leadership Collaborative Program through the Dartmouth Institute (15 month multidisciplinary quality improvement curriculum that includes a patient participant)
- Our nurse, respiratory therapist, and nurse practitioner participated in formal CF mentor programs
- We submitted grants to fund Physical Therapy and Pharmacy support in our clinic

We had a good showing at the **North American CF Conference in Denver** in October with attendance by many members of our multidisciplinary team. We were proud to present a poster and oral presentation on our CF Pulmonary Rehabilitation Program. We are among the first in the country to develop a customized program for people with CF and look forward to presenting more on our patient outcomes at next year's conference.

The hot topic this year at the conference was "**The Triple**" CFTR modulator (tezacaftor/ivacaftor/VX-659/445) for patients with one or two copies of the F508del mutation. Results of phase II trials showed significant improvement in FEV_1 and quality of life (*NEJM* 2018; 379:1599-1620). Early phase III results appear consistent. This could truly be a game changer for ~90% of CF patients and holds great promise to be FDA-approved within a year. We remain dedicated to caring for CF patients anywhere along their disease spectrum, from normal lung function to lung transplant.



Inova Adult CF Team (From left to right): Chris King, MD; Kim Palczynksi, RRT; Quyen Duong, RRT; Elizabeth Davies, SW;Lauren Marinak, NP; A. Whitney Brown, MD; Gerilynn Connors, RRT; Erin Lopynski, RD; Melissa Bowen, RN; Kareem Ahmad, MD (Not shown: Shambhu Aryal, MD; Jessica Chun, NP; Meg Fregoso, NP)

Please check out: www.inova.org/colleenstory

Pulmonary Hypertension Program (by Oksana Shlobin, MD.)

In 2014, our program was one of the first 6 programs to be accredited as a Comprehensive Care Center by the Pulmonary Hypertension Association (PHA). We underwent a re-accreditation visit in December 2018, and anticipate re-accreditation in early 2019. Dr. Oksana Shlobin continues to lead the program, while Dr. Christopher King acts as the Associate Medical Director of the program. Dr.Mitchell Psotka, an advanced heart disease cardiologist with experience in pulmonary hypertension, has joined the program, thus allowing us to provide multidisciplinary care to our patients. We have three full time pulmonary hypertension dedicated RN coordinators, under the leadership of Denise Lewis, RN. In addition, we have partnered with two experienced echocardiologists Drs. Pam Sears-Rogan and Joan Zhao who have particular interest in the assessment of right ventricular function using 3-dimensional echocardiography. Finally, we have engaged a thoracic radiologist, Dr. Melanie Atkins, to assist with interpretation of pulmonary vascular and cardiac specific imaging including V/Q scans, dual energy CT angiography, and cardiac MRI.

We now follow ~375 pulmonary hypertension patients of various etiologies. In addition to providing cutting edge clinical care, we remain committed to clinical research. Patients are offered an opportunity to participate in a variety of clinical trials: we currently have four active PH registries, four industry sponsored trials, including studies for Group 1 PAH, HFpEF-related pulmonary hypertension, sarcoidosis related PH and interstitial lung disease related PH with several more pharmaceutical RCTs for Group 1 PAH projected to start in the next quarter.

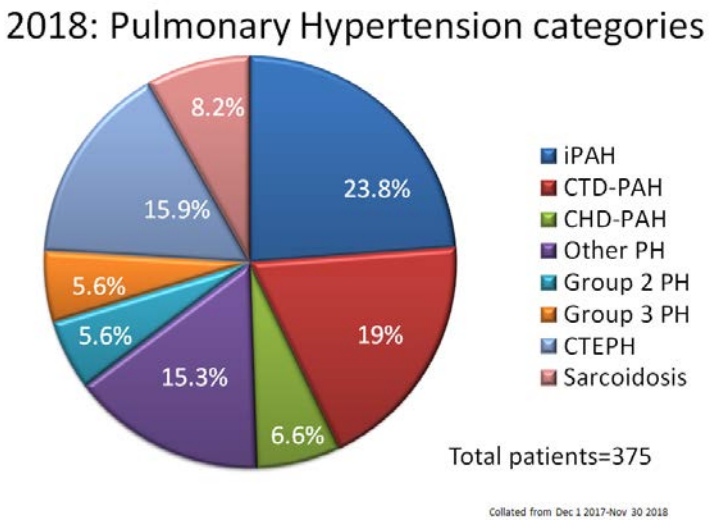
We continue to be active in outreach to increase awareness of this disease and provide education to both the medical and patient community. We hold a regional monthly multidisciplinary pulmonary hypertension meeting to discuss cases and provide didactic education on pulmonary hypertension topics. This conference is frequently attended by providers from esteemed Institutions including the NIH, Washington Hospital Center, and the University of Maryland. If you are interested in virtual or in-person participation, please contact Julieth Munoz at astrid.munoz@inova.org. Our first hospital wide Pulmonary Hypertension Education Day targeting nursing and midlevel staff was successfully held in the fall of this year with over 100 participants.

Left to right at the 1st Pulmonary Hypertension Education Day: Mitch Psotka MD, Oksana Shlobin MD, Linda Bogar MD, Christopher King MD. 2nd pic: Nargues Weir MD, Denise Lewis RN., Oksana Shlobin, MD.

Our team members continue to present at international, national and regional conferences including ISHLT, ATS, Chest, ERS, PHA and PHPN. Dr. Nathan had the honor of chairing a committee at the most recent World Symposium on Pulmonary Hypertension in Nice, France.

We welcome your referrals and are dedicated to partner with you in the care of this complicated patient population. Please do not hesitate to reach out to Dr.Oksana Shlobin (Oksana.shlobin@inova.org or (703) 776-2256) or Dr. Christopher King (Christopher.king@inova.org or (703) 776-4979). Referrals can be

directed to our dedicated intake coordinator Melany Vidaurre Llanos via email Melany.Vidaurrellanos@inova.org or fax (703) 776-8799).



Advanced Lung Disease & Lung Transplantation Critical Care (by Chris King, MD)

The capabilities of Inova Fairfax Hospital to care for the most critically ill patients continue to expand, particularly with regards to use of mechanical support. We have grown into a high volume extracorporeal membrane oxygenation (ECMO) center, with 78 ECMO "runs" in 2017 and on pace to perform 85 "runs" with over 18,000 hours of ECMO support in 2018. Over >90% of cases at INOVA are in the adult population, and about 1/3 are venovenous (VV) ECMO for respiratory failure. Comparative outcome data via the International ELSO registry to centers with similar patient volumes finds about 2 more patients surviving than expected (observed to expected survival ratio of 0.88 in all ECMO patients and 0.82 in VV ECMO patients). Referral for ECMO consideration can be initiated via the Cardiac Access center at 703-776-5905. Expansion of our ECMO program has translated into an increased ability to support our patients who are listed for transplantation as well as accept more complex patients for evaluation. In 2018, four patients were transplanted with ECMO support and patients have also received ECMO in the immediate post-transplant period. Two patients had severe pulmonary arterial hypertension and required awake cannulation to stabilize them prior to listing, a first for Inova Fairfax Hospital.

We have also extended our commitment to clinical research in the critical care/ ECMO arena. A new clinic to assess functional and psychological outcomes in ECMO survivors, a needed endeavor which is rarely available in most ECMO centers, is underway and will provide valuable information regarding short and long term function of these patients. Clinical research on methods to optimize anticoagulation monitoring and reduction in bleeding and thrombotic complications also continues.

Finally, we are a clinical trial site for the VENT-AVOID trial https://clinicaltrials.gov/ct2/show/NCT03255057) evaluating the A-LUNG machine in patients with COPD exacerbations. Patients failing non-invasive ventilation or failing to liberate from mechanical ventilation are eligible. This device is analogous to "lung dialysis", clearing CO2 through a low flow machine and a small venous cannula that can be placed at the bedside . If you have patients you wish to be considered for this trial please contact Dr. Christopher King at 703-953-7837.

Research (by A. Whitney Brown and Chris King)

The Advanced Lung Disease Research Program was established in 1996 and has grown exponentially since then. Our site participates in numerous clinical trials for a variety of lung diseases including interstitial lung disease, lung transplantation, pulmonary hypertension, chronic obstructive pulmonary disease, and non-CF bronchiectasis. This includes industry sponsored clinical trials, Inova investigator initiated studies, and research collaborations.

The research program's infrastructure includes:

- 4 research nurses
- 4 clinical research coordinators (CRCs)
- 1 research assistant
- 1 regulatory coordinator

Three of our research staff members (2 nurses/1 CRCs) are certified as Clinical Research Coordinators through the Association of Clinical Research Professionals (ACRP) and 1 CRC is certified as a Clinical Research Professional through Society of Clinical Research Associates (SOCRA).

Extensive experience with recruitment strategies in pulmonary trials as well as the tight integration of our clinical and research teams promotes effective communication with each other and patients. Every patient who is seen in our clinic is screened for available clinical trials on a daily basis by our research assistant. Our physicians personally discuss the importance and merits of clinical trial involvement, which raises patients' comfort and interest in participating. Our team's willingness to collaborate with sponsors and other institutions has led to exciting and novel studies.

Our research team has biweekly morning research team meetings to address upcoming trials, track enrollment in current trials and troubleshoot barriers to enrollment. In addition, the research team has dedicated weekly meeting times with the PI of all studies to review recruitment goals, progress of the study, and ensure all documentation is being completed appropriately and in a timely manner.

Our hospital is also home to a Clinical Trials Unit where most of our research patients are seen. The unit has two pulmonary function test rooms and a six-minute walk hallway, which allows the unit to serve as a one-stop shop for the majority of our research procedures. This unit can accommodate complex studies to include phase 1 trials, as well as overnight stays and multiple pharmacokinetic time points. With resources like this unit, our program continues to grow and aims to provide the best possible care for our patients.

This year, we would like to highlight…

Group 3 Pulmonary Hypertension

Inova Advanced Lung Disease and Transplant Clinic continues to be at the forefront of treatment of World Health Organization (WHO) Group 3 Pulmonary Hypertension. Dr. Nathan served as the Chair of the Task Force on PH due to Lung Diseases at the 6th World Symposium on Pulmonary Hypertension in Nice, France. Additionally, we have launched several trials studying novel therapies in Group 3 PH.

1. The INCREASE trial (https://clinicaltrials.gov/ct2/show/NCT02630316) is assessing the efficacy of inhaled treprostinil in Group 3 PH due to fibrotic lung disease. This study is currently enrolling and available to patients with combined pulmonary fibrosis and emphysema, a group excluded from most clinical trials.
2. The PERFECT study (https://clinicaltrials.gov/ct2/show/NCT03012646), also sponsored by United Therapeutics, is examining inhaled treprostinil in severe Group 3 PH due to COPD. Patients must have a mean pulmonary artery pressure ≥ 35 mmHg to qualify. Enrollment for this trial will be opening soon.
3. Finally, the PH-ILD study (https://clinicaltrials.gov/ct2/show/NCT03267108), sponsored by Bellarophon Therapeutics, is testing a novel inhaled nitric oxide delivery device in Group 3 PH due to interstitial lung disease.

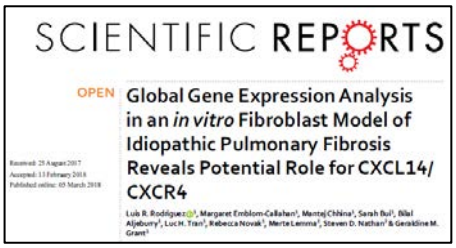
If you are interested in referring a patient to one of these trials, or have a patient with Group 3 PH who needs a clinical evaluation, feel free to contact Christopher King at christopher.king@inova.org or 703-953-

7837.

Academic Accomplishments 2018
(Inova authors bolded)

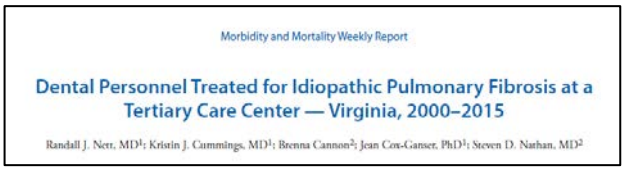
Original Research Manuscripts (accepted or published)

1. Agbor-Enoh S, Jackson A, Tunc I, Berry G, **Cochrane A**, Grimm D, Davis A, Shah P, **Brown AW**, Wang Y, Timofte I, **Shah P**, Gorham S, Wylie J, Goodwin N, Jang MK, Marishta A, Bhatti K, Fideli U, Yang Y, Luikart H, Piroozna M, Zhu J, Iacono A, **Nathan SD**, Orens J, Valantine HA, Khush K. Clinical antibody-mediated rejection (AMR) is a late manifestation of allo-antibody- associated injury. J Heart Lung Transplant. 2018 Jul;37(7):925-932.
2. Woolstenhulme JG, Guccione AA, Herrick JE, Collins JP, **Nathan SD**, Chan LE, Keyser RE. Left Ventricular Function Before and After Aerobic Exercise Training in Women Who Have Pulmonary Arterial Hypertension. Accepted to Journal of Cardiopulmonary Rehabilitation and Prevention. Accepted Oct 3 2018
3. Rodriguez LR, Emblom-Callahan M, Chhina M, Bui S, Aljeburry B, Tran LH, Novak R, **Nathan SD**, Grant GM. Novel Role for CXCL14/CXCR4 revealed through Global Gene Expression Analysis in an in vitro Fibroblast Model of Idiopathic Pulmonary Fibrosis. Sci Rep. 2018 Mar 5;8(1):3983. doi: 10.1038/s41598-018-21889-7.


SCIENTIFIC REPORTS

OPEN Global Gene Expression Analysis in an *in vitro* Fibroblast Model of Idiopathic Pulmonary Fibrosis Reveals Potential Role for CXCL14/CXCR4

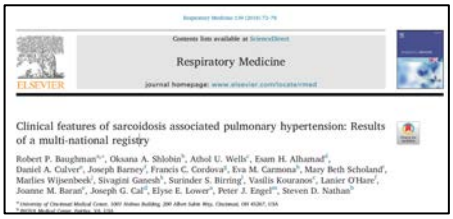
4. **Warren WA**, **Franco-Palacios D**, **King CS**, **Shlobin OA**, **Nathan SD**, **Katugaha S**, **Mani H**, **Brown AW.** A 24-year-old Woman with Precipitous Respiratory Failure Requiring Lung Transplantation. Chest 2018;153:e53-e56 (case report)
5. Yeo J, Chen T, Crawford EL, Zhang X, Khuder S, Levin A, Nana-Sinkam P, Massion PP, Arenberg DA, Midthun D, Mazzone PJ, **Nathan SD**, Wainz R, Silvestri G, Tita J, Willey JC. A COPD classifier based on targeted next generation sequencing analysis of genome maintenance genes in normal airway epithelial cells. BMC Pulm Med. 2018 Mar 5;18(1):42. doi: 10.1186/s12890-018-0603-y.
6. Nett RJ, Cummings KJ, **Cannon B**, Cox-Ganser J, **Nathan SD**. Dental Personnel Treated for Idiopathic Pulmonary Fibrosis at a Tertiary Care Center — Virginia, 2000–2015. MMWR Weekly March 9 2018;Vol. 67, No. 9


Morbidity and Mortality Weekly Report

Dental Personnel Treated for Idiopathic Pulmonary Fibrosis at a Tertiary Care Center — Virginia, 2000–2015

Randall J. Nett, MD[1]; Kristin J. Cummings, MD[1]; Brenna Cannon[2]; Jean Cox-Ganser, PhD[1]; Steven D. Nathan, MD[2]

7. Behr J, **Nathan SD**, Harari S, Wyuts W, Kirchgaessler K, Bengus M, Gilberg F, Wells A. Sildenafil added to pirfenidone in patients with advanced idiopathic pulmonary fibrosis and pulmonary hypertension: A Phase IIb randomised double-blind placebo-controlled study. Respir Med. 2018 May;138:13-20. doi: 10.1016/j.rmed.2018.03.019.
8. **King CS**, **Brown AW**, **Shlobin O**, **Weir N**, **Libre M**, **Franco-Palacios D**, **Ahmad S**, **Nathan SD**. Incidence and Impact of WHO Group 3 Pulmonary Hypertension in Idiopathic Nonspecific Interstitial Pneumonia. Eur Respir J 2018; 52: 1800545. doi: 10.1183/13993003.00545-2018
9. Raghu G, van den Blink B, Hamblin MJ, **Brown AW**, Golden JA, Ho LA, Wijsenbeek MS, Vasakova M, Pesci A, Antin-Ozerkis DE, Meyer KC, Kreuter M, Santin-Janin H, Mulder GJ, Bartholmai B, Gupta R, Richeldi L. Effect of Recombinant Human Pentraxin 2 vs Placebo on Change in Forced Vital Capacity in Patients With Idiopathic Pulmonary Fibrosis: A Randomized Clinical Trial. JAMA. 2018 Jun 12;319(22):2299-2307.
10. Baughman RP, **Shlobin OA**, Alhamad EH, Cal GJ, Porquero EC, Birring S, Lower EE, Engel PJ, **Nathan SD**. Clinical features of sarcoidosis associated pulmonary hypertension: results of a multi-national registry. Respir Med. 2018 Jun;139:72-78. doi: 10.1016/j.rmed.2018.04.015.



11. Maher TM, Swigris JJ, Kreuter M, Wijsenbeek M, Cassidy N, Ireland L, Axmann J, Nathan SD. Identifying barriers to IPF treatment- A survey of patient and physician views. Respiration epub Aug 18 2018 https://www.karger.com/Article/Abstract/490667.

12. Arun J, King CS, Shlobin OA, Brown AW, Wang C, Nathan SD. Pulmonary Hemodynamic Responses Predict Outcomes in Patients with Fibrotic Lung Disease Undergoing Exercise Right Heart Catheterization Testing. Eur Respir J 2018 52:1801015; published ahead of print 2018, doi:10.1183/13993003.01015-2018
13. Nathan SD, Lancaster LH, Albera C, Glassberg MK, Swigris JJ, Gilberg F, Kirchgaessler KU, Limb SL, Petzinger U, Noble PW. Dose modification and dose intensity during treatment with pirfenidone: analysis of pooled data from three multinational Phase III trials. BMJ Open Respir Res. 2018 Aug 2;5(1):e000323. doi: 10.1136/bmjresp-2018-000323. eCollection 2018.
14. Rhodes C et al. (Nathan SD among 118 co-authors). Genetic determinants of risk and survival in pulmonary arterial hypertension. Lancet Res Med Published online December 5, 2018 http://dx.doi.org/10.1016/S2213-2600(18)30409-0
15. Nathan SD, Costabel U, Glaspole I, Glassberg MK, Lancaster LH, Lederer DJ, Pereira CA, Trzaskoma B, Morgenthien E, Limb SL, Wells AU. Incidence of Multiple Progression Events: Pooled Analysis of Patients With Idiopathic Pulmonary Fibrosis Treated With Pirfenidone. accepted to Chest November 5th 2018
16. Raghu G, Flaherty KR, Lederer DJ, Lynch DA, Colby TV, Myers JL, Groshong SD, Larsen BT, Chung JH, Steele MP, Benzaquen S, Calero K, Case A, Criner G, Guerrero J, Nathan SD, Rai N, Ramaswamy M, Hagmeyer L, Davis JR, Gauhar U, Pankratz DG, Choi Y, Huang J, Walsh PS, Neville H, Lofaro LR, Barth NM, Kennedy GC, Brown KK, Martinez FJ. Diagnostic accuracy of conventional transbronchial biopsies through the use of a molecular classifier for usual interstitial pneumonia pattern. Accepted to Lancet Res Med 12/17/2018

<u>Reviews</u>

1. Brown AW, Nathan SD. The Value and Application of the Six Minute Walk Test in Idiopathic Pulmonary Fibrosis. Ann Am Thorac Soc. 2018 Jan;15(1):3-10.

2. Benza R, Raina A, Kanwar MK, Nathan SD, Mathai SC. sGC stimulators: evidence in pulmonary hypertension and beyond. accepted to the Journal of Rare Diseases Research and Treatment 11/2/17.
3. Aryal S, Nathan SD. An Update on Emerging Drugs for the Treatment of Idiopathic Pulmonary Fibrosis. Expert Opinion on Emerging Drugs. Expert Opin Emerg Drugs. 2018 Jun;23(2):159-172. doi: 10.1080/14728214.2018.1471465.
4. Aryal S, Nathan SD. Single vs Bilateral Lung Transplantation: When and Why. Curr Opin Organ Transplant. 2018 Jun;23(3):316-323. doi: 10.1097/MOT.0000000000000527.

5. El-Chemaly S, O'Brien KJ, Nathan SD, Weinhouse GL, Goldberg HJ, Connors JM, CuiY, Astor TL, Camp PC, Rosas IO, WoldeHanna ML, Speransky V, Merideth MA, Gahl WA, Gochuico BR. Clinical management and outcomes of patients with Hermansky-Pudlak syndrome pulmonary fibrosis evaluated for lung transplantation. PLoS One. 2018 Mar 16;13(3):e0194193. doi: 10.1371/journal.pone.0194193. eCollection 2018.
6. King CS, Ryan L, Singh R. VA ECMO Review. Crit Care Clinics.
7. King CS, Brown AW, Aryal S, Ahmad K, Donaldson S. Critical Care of the Adult Cystic Fibrosis Patient. Chest. 2018; pii: S0012-3692(18)31123-1. doi: 10.1016/j.chest.2018.07.025. [Epub ahead of print].
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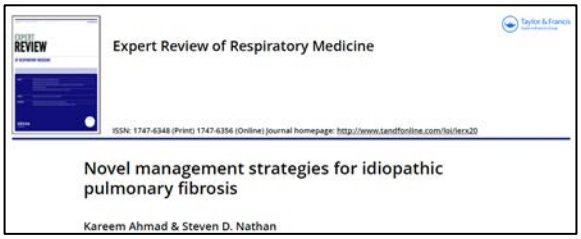


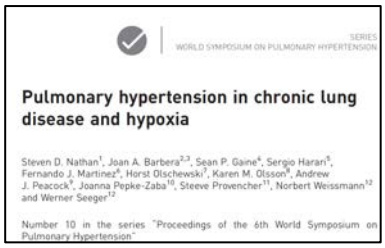
9. King CS, Shlobin OA. Ask the Expert: Thyroid Disease in PAH. Advances in Pulm HTN. Submitted

Editorial

Brown AW. Unclassifiable Interstitial Lung Disease: Time to Shrink the Black Box. Annals ATS 2018; 15(7):806–807.

Consensus Statements

1. Wells AU, Poletti V, Behr J, Cassidy N, Costable U, Cottin V, Hansell DM, Masefield SC, Richeldi L, Ross D, Ancochea J, Antoniou KM< Bajwah S, Bouros D, Brown KK, Collard HR, Corte TJ, Crestani B, Dai H, Drent M, Egan JJ, Fell CD, Fischer A, Flaherty KR, Grutters JC, Hirani N, Inoue Y, Maher TM, Muller-Quernheim J, Nathan SD, Noble PW, Powell P, Robalo-Cordeiro C, Ryerson CJ, Ryu JH, Saltini C, Selman M, Sverzellati N, Taniguchi H, Undurraga A, Valeyre D, Vancheri C, Wuyts W, Xaubet A.Diagnosis and management of idiopathic pulmonary fibrosis - a combined physician and patient European Respiratory Society and European Lung Foundation consensus statement. Accepted Eur Res J May 14th, 2018
2. Nathan SD, Barbera JA,Gaine SP, Harari S, Martinez FJ, Olschewski H, Olsson KM, Peacock AJ, Pepke-Zaba J, Provencher S, Weissmann N, Seeger W. Pulmonary Hypertension in Chronic Lung Disease and Hypoxia. Accepted to ERJ 10/11/2018



Case Reports

1. Jose A, Franco-Palacios D, King CS, Gomberg-Maitland M. A 70 year old woman presenting with diarrhea and in-hospital cardiac arrest. Chest 2018. 153(1): e5-e8.
2. King C, Clement S, Katugaha S, Brown AW. Fungal Thyroiditis in a lung transplant recipient. BMJ Case Reports 2018;doi:10.1136/bcr-2018-227033

Abstracts/Presentations
International Society for Heart and Lung Transplantation, April
Nice France 2018

1. Shah P, Rohly M, Joshi A, Timofte I, **Brown AW**, Orens J, Iacono A, **Nathan SD**, Avery R, Agbor-Enoh S, Valantine H. Donor Derived Cell Free DNA is elevated with pathogens that are risk factors for Chronic Lung Allograft Dysfunction.
2. Agbor-Enoh S, Jackson AM, Berry G, **Cochrane AB**, Tunc I, Jang M, Bhatti K, Marishta A, Gorham S, Yang Y, Fideli U, Zhu J, Pirooznia M, Grimm D, Luikart H, Shah P, Timofte I, Iacono A, Cao Z, **Brown AW**, Orens J, Marboe C, **Nathan SD**, Khush K, Valantine H. Antibody-mediated rejection: should we wait for clinical diagnosis?
3. **Brown AW**, Agbor-Enoh S, Shah P, Timofte I, Orens J, Iacono A, Lemme M, **Barnett S, Soares F, Nathan SD, Ahmad K**, Valantine H. Role of dd-cfDNA in Predicting Early Post-Operative Course in Lung Transplant Recipients.
4. Agbor-Enoh S, **Cochrane AB**, Jackson AM, I. Tunc, P. Shah, Z. Cao, **Brown AW**, Timofte I, Marishta A, Jang M, Gorham S, Yang Y, Fideli U, Iacono A, **Nathan SD**, Orens J, Valantine H. Is the New ISHLT Criteria for Pulmonary Antibody-Mediated Rejection also a Severity Indicator?
5. Agbor-Enoh S, Tunc I, Gorham S, Jang M, Fideli U, Marishta A, Zhu J, Pirooznia M, Yang Y, Davis A, Grimm D, Cao Z, Luikart H, Shah P, Timofte I, **Brown AW**, Iacono A, **Nathan SD**, Orens J, Khush K, Valantine H. Clinically-unrecognized allograft injury is common after lung transplantation.
6. **Ahmad K, Brown AW**, Shah P, Timofte I, Orens J, Iacono A, Lemme M, **Aryal S**, Cochrane A, **Nathan SD**, Agbor-Enoh S, Valantine H. Role of dd-cfDNA in Predicting Early Hospital Readmission in Lung Transplant Recipients.
7. Jackson AM, **Cochrane AB, Nathan SD, Brown AW**, Shah P, Orens J, Timofte I, Pham SM, Gorham SS, Bhatti K, Marishta A, Jang M, Yang Y, Fideli U, Tunc I, Zhu J, Iacono I, Agbor-Enoh S, Valantine H. Elevated Donor-Derived Cell-Free DNA (ddcfDNA) as an early risk factor for the development and persistence of de novo donor specific HLA antibody.
8. **Bogar L, Chun J, Fregoso M, Cantwell L, Shah P**. Negative Pressure Wound Therapy Applied to Groin Cannulation Incisions Decrease the Incidence and Severity of Seroma Formation.
9. **Nunes FS, King CS, Nathan SD, Fregoso M, Ahmad K, Aryal S, Brown AW, Barnett SD, Shlobin OA, Vester A.** High Body Mass Index is a Risk Factor for Acute Cellular Rejection in Lung Transplant Recipients.
10. **Bowen MA, Cochrane AB, Albergottie-Barnes L, Brown AW**. Rapid High Dose Vaccine Series: Increasing Immunity to Hepatitis B in Lung Transplant Candidates.
11. Jose A, **King C**, Welt E, **Shlobin OA, Brown AW, Aryal S, Weir N, Nathan SD**. Abnormal Invasive Pulmonary Hemodynamics Predict Outcomes in Exercising Patients.
12. Jarrett H, Jonnalagadda AK, Liu SD, Bagnola AJ, Lewis D, Shlobin OA, Barnett CF. Rapid Transition from Parenteral to Oral Treprostinil in PH is Feasible and Safe: A Retrospective Cohort Study.

<u>American Thoracic Society May, San Diego, CA</u>

1. Nett RJ, Cummings KJ, **Cannon B**, Cox-Ganser J, **Nathan SD**. Dental Personnel Treated for Idiopathic Pulmonary Fibrosis at a Specialty Clinic.
2. Barochia AV, Kaler M, Gordon EM, Figueroa D, **Weir NA, Lemma WoldeHanna M**, Sampson M, Remaley AT, **Barnett SD**, Grant G, **Nathan SD**, Levine SJ. Serum Lipids and Lipoproteins Quantified by NMR Spectroscopy are Correlated with FVC, 6MWD and GAP index in Idiopathic Pulmonary Fibrosis.
3. Waxman AB, Tapson VF, Smith PM, Deng C, **Nathan SD**. A Multicenter, Randomized, Double-Blinded, Placebo-Controlled Trial to Evaluate the Safety and Efficacy of Inhaled Treprostinil in Subjects with Pulmonary Hypertension due to Parenchymal Lung Disease (Study RIN-PH-201).
4. Behr J, **Nathan SD**, Harari S, Wuyts W, Kirchgaessler K, Bengus M, Gilberg F, Wells A. Baseline Characteristics From a Pre-specified Interim Analysis of a Phase IIb, Randomized, Double-Blind, Placebo-Controlled Trial of Sildenafil Added to Pirfenidone in Patients With Advanced Idiopathic Pulmonary Fibrosis and Pulmonary Hypertension.
5. **Poreddy M, Nunes FS, Scully A, Nathan SD, Brown AW, Ahmad K, King CS, Shlobin OA, Aryal S, Weir N**. Gender and BMI Predict Antifibrotic Tolerance in IPF.
6. **Chun J, King C, Fregoso M, Marinak L, Shlobin O, Aryal S, Nathan SD, Brown AW**. Incidence and Impact of Gallbladder Disease after Lung Transplantation.
7. Jose A, **King C**, Welt E, **Shlobin OA, Brown AW, Aryal S, Weir N, Nathan SD**. Abnormal Exercise Pulmonary Hemodynamics Predict Outcomes in Patients with Lung Disease.
8. Biru N, **King CS, Shlobin OA, Aryal S, Nathan SD, Marinak L**, Woods C, **Brown AW, King CS**. Rare case of rapidly progressive interstitial lung disease following adult tetanus, diptheria, and pertussis (Tdap) vaccination.
9. Mabe D, **Shlobin OA, Bogar L, Nathan SD, Brown AW, Aryal S, Murphy C, King C**. ECMO as a Bridge to Initial Medical Therapy in a Patient with Decompensated Pulmonary Arterial Hypertension. Submitted ATS 2018 OS to FU
10. **Nunes FS, Vester A, Brown AW, King CS, Ahmad K, Fregoso M, Aryal S, Nathan SD, Barnett SD, Shlobin OA.** Body Mass Index at Listing May Impact Survival After Lung Transplantation.
11. **Ewarien B, Brown AW, Libre M, Cochrane A, Marinak L, Ghandi B, Shlobin OA, Nathan SD, King CS**. Paravertebral Nerve Block For Pain Control Following Lung Transplantation.
12. Kouranos V, **Shlobin OA, Nathan S**, Wells A, Baughman RP. Factors associated with reduced survival in sarcoidosis associated pulmonary hypertension: results of the registry for sarcoidosis associated pulmonary hypertension (ReSAPH).
13. **Cheng JM, Soares F, Cannon B, Brown AW, Nathan SD**. The White Blood Cell Count as a Prognostic Indicator in Idiopathic Pulmonary Fibrosis.
14. Gersten RA, **Cannon B, Bowen M, Davies E, Brown AW**. Evaluation of Depression and Anxiety and their Influence on Outcomes in Adult Patients with Cystic Fibrosis.

<u>World Symposium on Pulmonary Hypertension Nice, France Feb 2018</u>

Behr J, **Nathan SD**, Harari S, Wuyts W, Kirchgaessler K, Bengus M, Gilberg F, Wells A. Sildenafil added to pirfenidone in patients with advanced idiopathic pulmonary fibrosis (IPF) and pulmonary hypertension (PH): a Phase IIb, randomized, double-blind, placebo-controlled study. Presented at World PH meeting Nice, France 2018

<u>American Society for Echocardiography 2018</u>

Benedict C, **Shlobin O**, Shiao NJ, Welt E, Vakilzadeh M, **Nathan SD, Zhao Q**. Characterization of Left Ventricular Diastolic Function Difference in Patients with Exercise-induced Pulmonary Hypertension due to Heart Failure with Preserved Ejection Fraction vs Pulmonary Arterial Hypertension.

<u>European Respiratory Society 2018</u>

K.R. Flaherty, J.A. de Andrade, L.H. Lancaster, K.O. Lindell, **S.D. Nathan**, G. Raghu, C. Spino, J.L. Stauffer, P.J. Wolters, G.P. Cosgrove Baseline Characteristics of the Initial 1461 Participants in the Pulmonary Fibrosis Foundation. To be submitted to ERS 2018

<u>Chest San Antonio, TX October 2018</u>

1. **Nathan SD**, Costabel U, Albera C, Behr J, Wuyts WA, Kirchgaessler K, Stauffer JL, Morgenthien E, Yang M, Limb SL, Noble PW. Effect of Pirfenidone on

Exercise Capacity and Dyspnea in Patients with Idiopathic Pulmonary Fibrosis and More Advanced Lung Function Impairment. Ankush Ratwani, MD;

2. Kareem Ahmad, MD; Christopher S. King, MD; Oksana Shlobin, MD; Nargues Weir, MD; Shambhu Aryal, MD; Steven D. Nathan, MD; A. Whitney Brown, MD. Connective Tissue Disease associated Interstitial Lung Disease and Outcomes after Hospitalization: a Cohort Study. Podium Presentation
3. Ahmad K, Shlobin O, Nathan S, King CS, Aryal S, Brown AW. Rising Incidence of Pulmonary Embolism Post-Lung Transplantation: A Single Center Experience. Submitted to Chest 2018.

North America Cystic Fibrosis Conference 2018

Bowen M, Duong Q, Russell C, Lamberti J, Connors G, Brown AW. Development and implementation of a CF specific outpatient pulmonary rehabilitation program. Podium presentation and poster

America Society for Matrix Biology 2018

Bui S, Young O, Nathan SD, Paige M., Grant GM. Novel Tacrolimus (FK506) Analogue Inhibits FKBP10:Lysyl-Hydroxylase-2 (LH2) Collagen Processing in Idiopathic Pulmonary Fibrosis (IPF) Fibroblasts. American Society for Matrix Biology, October 14-17, 2018.

Patient Education & Support Groups

Transplantation. We hold monthly transplant education and support group to which all our pre-transplant and post-transplant patients are welcome. It is an expectation that our listed patients attend this as it also functions as an education forum with specific topics and speakers on a monthly basis. This year we hope to institute webinars of this for our listed patients who live more than 2 hours away.

IPF. The Pulmonary Fibrosis Support Group of Metropolitan Washington DC is a monthly forum for not only IPF patients, but also those patients with any form of pulmonary fibrosis or interstitial lung disease. Patients do not have to be our clinic patients in order to attend. This support group is now supported by the Pulmonary Fibrosis Foundation and takes place the 4th Tuesday of every month at 1pm in the Physician Conference Center lower level at Inova Fairfax Hospital (see flyer)

Pulmonary Hypertension. We also have a patient run PH support group for all patients with any form of pulmonary hypertension. There are two Pulmonary Hypertension support groups in the area; one in Virginia (NOVA@PHASupportGroups.org) and one in Maryland (MD-SouthernMD@PHASupportGroups.org)

For any information pertaining to our Support Groups, please contact our Social Worker, Susan Perry at susan.perry@inova.org

Agenda for Education Group
Heart and Lung Transplantation
Ventricular Assist Device, VAD
Inova Fairfax Hospital: Heart and Vascular Institute
2019

January 8	**Pulmonary & Cardiac Rehab**	**Rehab Staff**
February 5	**Cardiac Transplant** VAD Boot Camp to Follow	**Shashank Desai, MD**
March 5	**Lung Transplant**	**A. Whitney Brown, MD**
April 2	**Post Transplant Care**	**ICU/3rd Floor Nurses**
May 7	**Dermatology**	**Jennifer DeSimone, MD**
June 4	**Clinic and Meds**	**Post Nurse Practitioners**
July 2	**Life Planning**	**Social Workers/Palliative Care**
August 6	**Living With a Transplant** VAD Boot Camp to Follow	**Patient Panel**
September 3	**Waiting for a Transplant**	**WRTC Organ Procurement Org.**
October 1	**Health Insurance Issues**	**Financial Coordinators**
November 5	**Nutrition**	**Leah Papazian & Elise Kouri**
December 3	**Infectious Disease**	**Shalika Basnayake Katugaha,MD**

First Tuesday of Every Month from 1:00 to 2:00 p.m.
IHVI Conference Center- Ground floor, VAD patients to meet 2:00-2:30
*** All listed candidates need to call (703) 776-7076 if unable to attend**

PULMONARY FIBROSIS SUPPORT GROUP
of Greater Washington DC
WWW.PF-VA.ORG

MEETINGS
TIME: 1:00 PM TO 2:30 PM

Come early, lunch and socialize with the other patients & caregivers (We'll be in the room at 12:00)

Website: www.pf-va.org – Facebook: look for Pulmonary Fibrosis Support Group of Greater Washington DC

RSVP to: 703-776-5776 / Susan.Perry@inova.org

- Please advise in advance of any oxygen requirements.
- We are open to all PF patients, family members, caregivers, medical professionals and suppliers. *You do not need to be a patient at Inova Fairfax to attend.*

PULMONARY FIBROSIS SUPPORT GROUP of Greater Washington DC

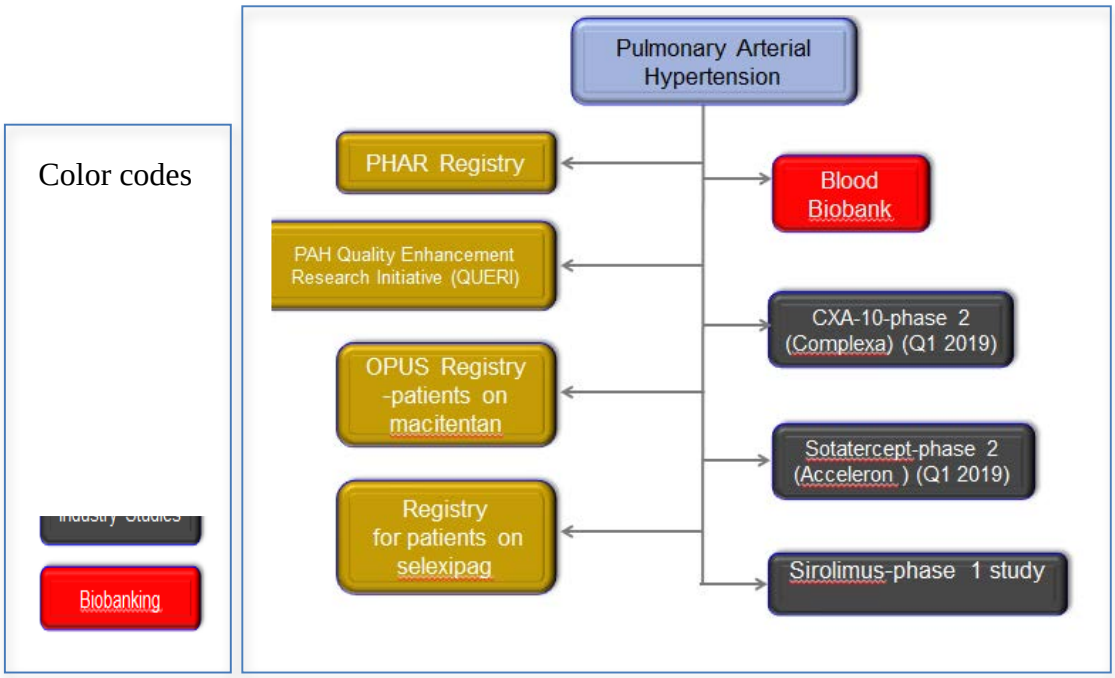
Meetings
4th Tuesday of each Month in 2019

January 22
February 26
March 26
April 23
May 28
June 25
July 23
August 27
September 24
October 22
November 26

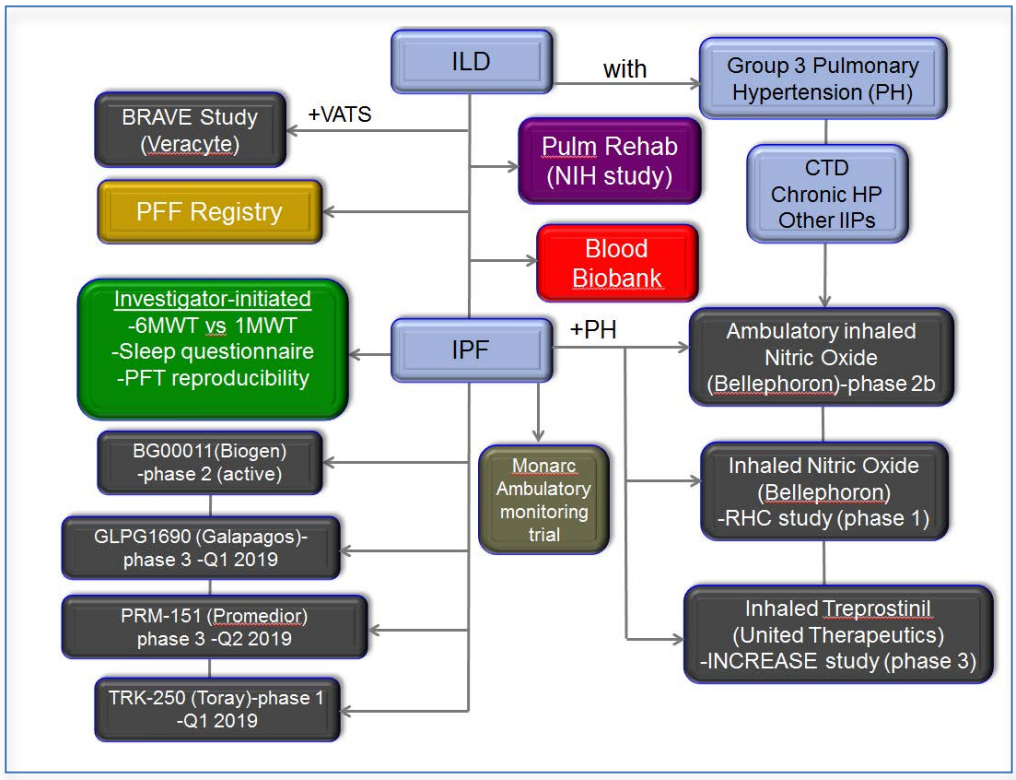
INOVA FAIRFAX HOSPITAL
3300 GALLOWS ROAD
FAIRFAX, VA 22042
Park in the **BLUE** Garage. Come to the Physicians Conference Center - across the street to the right. You will be guided from there!!

Currently Available Research Studies
for all inquiries please email:
lungresearch@inova.org

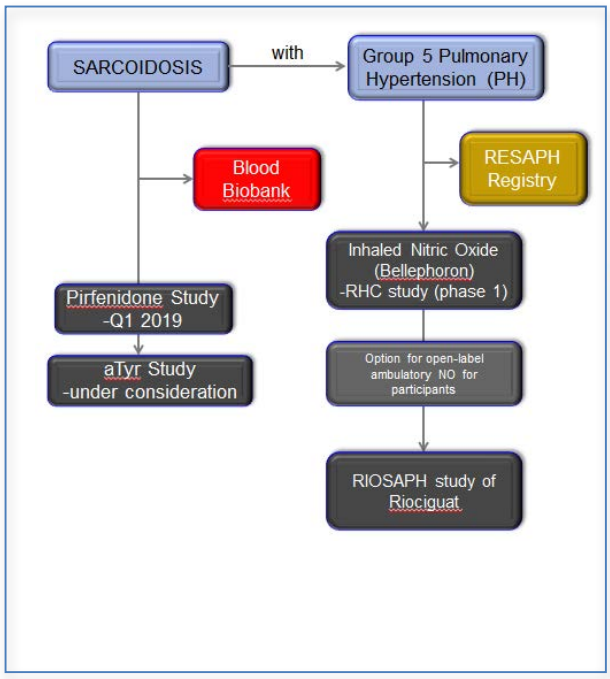
Clinical Trial algorithm for Pulmonary Arterial Hypertension



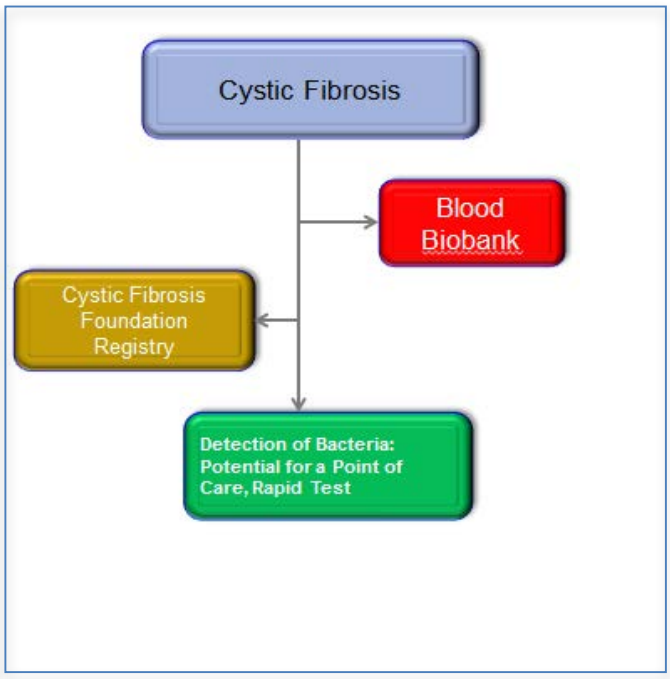
Clinical Trial algorithm for Interstitial Lung Disease



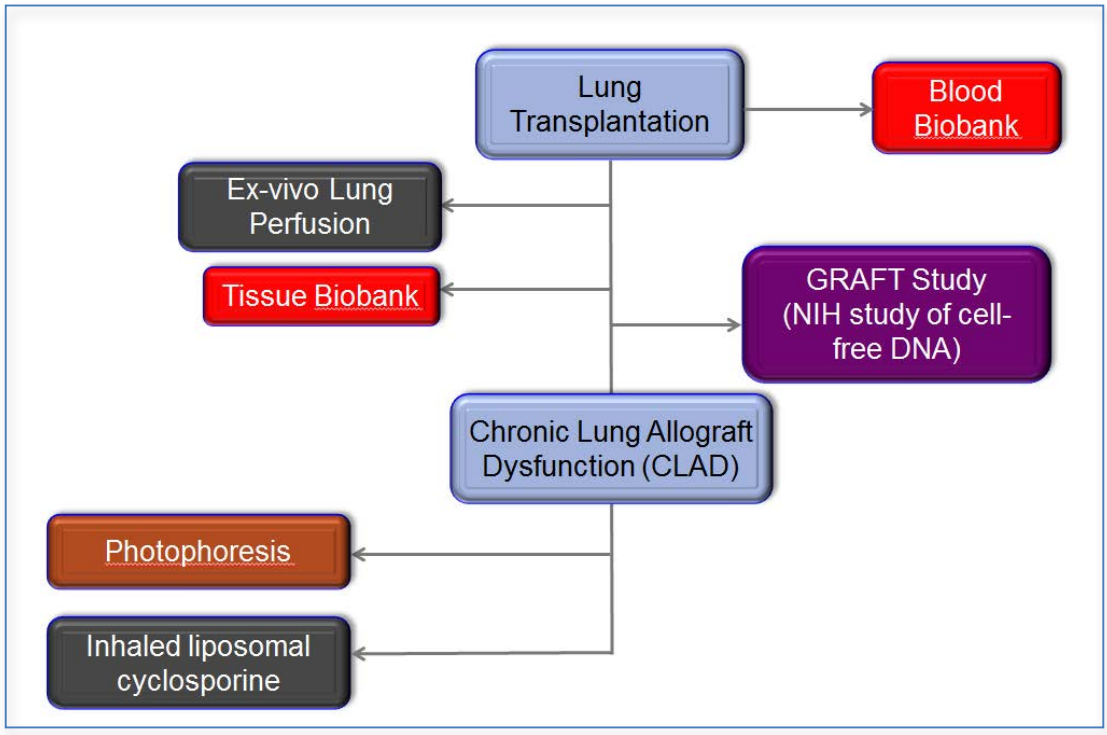
Clinical Trial algorithm for Sarcoidosis



Clinical Trial algorithm for Cystic Fibrosis



Clinical Trial algorithm for Lung Transplantation



Inova Lung Transplant and Advanced Lung Disease Team Members (December 2018)

Fourth row: Andrea Grajeda (Referral Coordinator); Melany Vidaurre-Llanos (PH administrative Assistant); Denise Lewis (PH and Lead Nurse Coordinator); Priscilla Dauphin (Research Coordinator); Serina Zorrilla (Research Coordinator); Quyen Duong (CF RT); Kim Auguste (Medical Assistant); Latoya Albergottie-Barnes (Nurse Coordinator).
Third row: Matthew Kott (Nurse Coordinator); Rodrick Likonko (Financial Coordinator); Sarah Scott (Office Manager); Jennifer Pluhacek (Research Coordinator); Brenna Cannon (Research Coordinator); Lori Hill (Financial Coordinator); Danielle Dacosta (Research Coordinator); Michelle Schreffler (Nurse Coordinator); Merte Lemma (Research Coordinator); Carlos Coronel (Sr. Admin Coordinator); Mathew Koslow (Advanced Lung Disease Fellow).
Second row: Adam Cochrane (Transplant Pharmacist); Jessica Chun (Nurse Practitioner); Leah Papazian (Dietician); Elizabeth Davies (Social Worker); Susan Perry (Social Worker); Deanna Ridgeway (Financial Coordinator); Tina Thronson (Quality Manager); Edwinia Battle (Research Manager); Erin Lopynski (Dietician); Astrid "Julieth" Munoz (Program Manager); Lauren Marinak (Nurse Practitioner); Meg Fregoso (Nurse Practitioner).
First Row: Shambhu Aryal, MD, Kareem Ahmad, MD; Nargues Weir, MD; Steven Nathan, MD; Oksana Shlobin, MD; Whitney Brown, MD; Shalika Katugaha, MD (Infectious Disease); Melissa Bowen (CF Coordinator); Debbie Campbell (Transplant Director).
Absent:
Interventional Pulmonologist: Amit "Bobby" Mahajan, MD
Surgeons: Linda Bogar, MD; Sandeep Khandhar, MD; Liam Ryan, MD; Eric Sarin, MD; Ramesh Singh, MD.
Pulmonologists: Chris King, MD; Osman Malik, MD.
Research: Lori Schlegel (Research Coordinator); Drew Venuto (Research Coordinator).
Nurse Coordinators: Karen Brown (PH Coordinator); Angela Scully (ALD Coordinator).