December 18, 2017

CTS INSPIRATIONS

CTS NEWS

President’s Letter

Dear colleagues and friends,

2017 has been an important and eventful year for CTS. I am grateful to all of you for your support and involvement in many of CTS’ activities and to our leadership and committees for your important contributions. A few areas that I am very grateful for are included below.

CTS’ greatest resource is our members. We thank you for your tireless and important work to improve the clinical care and science of pulmonary medicine, patient outcomes and meet the needs of the community. The response to the Sonoma – Napa and Southern California fires with rapid response, triage and care of countless patients is one important example of the bravery and skill that makes a difference where and when needed. We are deeply saddened by those who lost so much and suffered during these tragic times. For CTS resources, please click on the following link: https://calthoracic.org/2017fires/

We have also benefited from our outstanding administrative leadership team lead by Phil Porte, and colleagues Vickie Parshall, Karen Lui and Dave Eubanks. They have provided needed organizational stability and growth, and have skillfully overseen both our successful UCSD conference and the planning for our upcoming Carmel meeting on Friday January 26 and Saturday January 27, 2018. To register go to: https://calthoracic.org/events/2018-annual-educational-conference/.

Thank you to our professional leadership and committees including Angela Wang, immediate past president, George Chaux, treasurer, Philippe Montgrain, president elect, and committee chairs and co-chairs Philippe Montgrain, Shazia Jamil, Lorri Leard (Education), Trina Limberg, Bill Stringer, (Multidisciplinary), DJ Kaley (Pediatrics), George Chaux, Jeff Kupperman (membership), Jim Brown (Advocacy), Russell Klein (Clinical Programs) and our most recent committee chair Tisha Wang (Nominating). Thank you for all you do to enable accomplishment of our mission to improve California lung health and, through advocacy and education, advance the science and practice of pulmonary and critical care medicine.

We are grateful to our partner societies including California Society for Respiratory Care lead by Mike Madison with help from CTS board liaison Rick Ford, American Lung Association of California, the National Association of Medical Directors of Respiratory Care, California Society for Pulmonary Rehabilitation, and Rick Robbins for sharing the Southwest Journal of Pulmonary and Critical Care.
We are looking ahead to another exciting CTS conference at the beautiful Quail Lodge resort in Carmel on Friday January 26 and Saturday January 27, 2018. Friday’s symposium will feature a Multidisciplinary Approach to Interstitial Lung Disease featuring top international speakers, followed by fellow’s poster competition, reception and the annual Western Respiratory Club dinner featuring John Balmes MD speaking on ‘The Sonoma-Napa Fires: Health Impact and Response’. Saturday’s agenda includes a half day Lung Cancer Symposium followed by an Update on Bronchiectasis. For more information and to register, visit: https://calthoracic.org/events/2018-annual-educational-conference/.

Lastly, we are holding our 4th annual CTS fellow/multidisciplinary poster competition and there is still time to submit by January 1, 2018. It is open to trainees, faculty and multidisciplinary colleagues. For more information on our 2018 poster competition and to submit your abstracts on-line visit: https://calthoracic.org/2018-poster-abstract-submission/.

HAPPY HOLIDAYS TO ALL!!

Diagnosis of Pulmonary Hypertension in the Real World

By Sachin Gupta, MD FCCP and Dana McGlothlin, MD

The evaluation of pulmonary hypertension (PH) is anchored on determining the etiology, as management and prognosis vary significantly based on the type of underlying disease.

The key questions to be asked when evaluating someone with suspected PH are: Where is the lesion that is causing the elevated pulmonary vascular resistance? Is it post-capillary PH due to left heart disease, whether pure pulmonary venous hypertension or combined post- and pre-capillary PH (WHO Group 2 PH)? Is it pre-capillary PH, in which case the PH may be due to pulmonary arterial hypertension (WHO Group 1 PH)? The last 3 categories are important to consider as well: is the PH due to lung disease and/or hypoxemia (WHO Group 3 PH), due to chronic thromboembolic disease (CTEPH; WHO Group 4 PH), or due to miscellaneous diseases such as Sarcoidosis with unclear or multifactorial mechanisms (WHO Group 5 PH).

Having a clear picture is key for explaining to patients the necessary life-style changes, prognosis, and therapeutic management aspects of their disease.

The Kaiser Permanente Northern California PH cohort has provided insights into the real world care of PH. In our review, care is frequently provided with due diligence to the workup and with careful consideration of the role of chronic lung or left heart disease on the development of PH. Transplant evaluation, too, is sought appropriately in patients with both ILD and PH. However, as others have found, pitfalls sometimes arise in the diagnosis of WHO Group 1 PAH that can lead to inappropriate pulmonary vasodilator initiation and the resultant potential consequences (pulmonary edema, V/Q mismatch, hypotension, and of course tremendous expense without demonstrated benefit).
Key Points:

1) PH should be suspected based on symptoms, exam findings, and an Echocardiogram with a PASP estimate of >40 mmHg, especially when other features of PH are present. Other features suggestive of PH on echocardiography including RV enlargement and/or interventricular septal flattening. PH may still exist with a “normal” estimated PASP on echo when other features of PH are present.

2) The vast majority of patients with pre-capillary PH (including PAH) have an “impaired relaxation” type of left ventricular diastolic filling pattern (aka “mild” or “grade 1” diastolic dysfunction). Whereas the presence of any degree of left atrial enlargement and/or moderate to severe diastolic dysfunction (aka “grade 2” or higher diastolic dysfunction; or “pseudonormal” or “restrictive” diastolic filling pattern) strongly suggests post-capillary PH (i.e. WHO Group 2 PH).

3) Right heart catheterization (RHC) is not required all patients with PH on Echo, especially in patients with PH in the setting of severe lung disease or left heart disease when it is unlikely to change management.

4) RHC is absolutely necessary for the diagnosis of pulmonary arterial hypertension (PAH), in order to establish the diagnosis, exclude post-capillary PH etiology, screen for congenital shunt lesions with a saturation run, and to assess hemodynamics of prognostic significance (right atrial pressure and cardiac output) which help to guide initial PAH therapy decisions. No cath, no PAH therapy!

5) During RHC, obtaining a PA wedge/occlusion pressure (PCWP/PAOP) can be challenging and studies bear out large variations in accuracy. Therefore, obtaining a directly measured left ventricular end-diastolic pressure (LVEDP) can be helpful when it is difficult to obtain an accurate PCWP, or when the PCWP is counter to what is expected clinically.

6) Based on long term outcomes data, acute vasodilator testing is most useful and therefore indicated in truly idiopathic PAH and anorexigen induced PAH.

7) Ventilation/perfusion scanning is more sensitive than CT-Angiogram for excluding chronic PE as the etiology of the PH and is recommended first line for all patients undergoing a PH workup.

8) Understanding the risk factor in development of PH is critical, particularly in an era in which clinical trials are available (for example in Scleroderma, Sarcoidosis, and even ILD associated PH).

9) Finally, illicit substance abuse can be a challenge to identify, and less obvious in a highly functional patient. Urine drug toxicity screening should be regularly performed during the evaluation of PH (and more than once if amphetamine use is suspected clinically).

PH diagnosis can be challenging as often more than one potential etiology is identified. Assistance with PH diagnosis and management should be readily sought from a PAH Center when possible.
References:


## Southwestern Journal of Pulmonary and Critical Care Medicine

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