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EDITORIAL

Right Heart Catheterization—To Do or Not To Do? Introducing a New Diagnostic Algorithm for Pulmonary Hypertension

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The work up for pulmonary hypertension (PH) is exhaustive, expensive, and time consuming.[1,2] One look at the diagnostic algorithm and all that it entails can discourage even the most motivated clinician and creates an unreasonable burden on patients to undergo tests, many of which are of questionable necessity. It is difficult for people of limited means to coordinate work schedules and arrange finances to undergo the series of tests, thereby contributing to delayed diagnosis and worsened outcomes.[3] In addition, there are certain nuances to interpretations of the current recommended diagnostic tests that adds to the complexity. Ventilation/perfusion scan, used to identify Group 4, thromboembolic PH, can be difficult to interpret and is not available at smaller hospitals. In fact, over the past 2 years hospitals with capacity could not perform the ventilation component due to concerns regarding transmission of COVID-19.[4] Computed tomography of the chest requires expertise in looking at the peripheral vascular tree, the right ventricle, and potential congenital abnormalities. Even the "gold standard" right heart catheterization (RHC) has idiosyncrasies stemming from reliance on computer-generated averages and not end-expiratory measures.[5] Physicians' understanding of when to conduct and how to interpret reversibility testing, a fluid challenge, or exercise testing is lacking. Finally, when PH guidelines are written, as with all medical guidelines we take great pride as a group of experts to ensure that anything we support or endorse meets as high a level of evidence as possible: evidence-based medicine (most typically a double-blind randomized controlled trial). This high bar is expected but may not be appropriate for a rare disease.

See Article by Jansen et al.

Why then, when it comes to the diagnostic work up for PH, do we throw away those requirements and endorse algorithms that have never been evaluated or validated systematically? In this issue of the *Journal of the American Heart Association (JAHA)*, a paper by Jansen and colleagues describes an inadequate workup of patients with PH by virtue of omission of RHC in community hospitals in the Netherlands. The authors as part of the PH group/consortium in the Netherlands sought to determine if appropriate care is occurring at community hospitals with respect to PH evaluation and diagnosis.[6] The consortium used their data set built from 12 hospitals to determine if patients had a RHC and the reasons for omission. The authors quite reasonably argue that, by arbitrarily omitting hemodynamic assessment in patients during their PH workup, patients with Group 2 PH, have delays in treatment. Without a RHC pulmonary, seen by leading

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US PH centers, pulmonary arterial hypertension patients may be missed and misassigned as (most likely) pulmonary venous hypertension.[7]

One justification as seen by Jansen and colleagues for not doing a RHC, early in the diagnosis is the inaccessibility of the procedure at the hospital and hesitation in performing the RHC in patients with multiple comorbidities.[6] Advanced age and echocardiographic parameters indicative of left heart disease were associated with **not** having a RHC, whereas the presence of prior thromboembolism or pulmonary arterial hypertension-associated conditions, right atrial dilation, and severe tricuspid regurgitation on echocardiography influenced physicians to perform the RHC. Cost to the patient also may influence the decision. A quick tabulation of cost by the current algorithm versus a RHC done first promptly dismisses this argument. The national average for echocardiogram is $1684. If we then follow the 2009 American College of Cardiology Foundation/American Heart Association document, the national average of a ventilation-perfusion scan is $1569. The national average of a pulmonary function test is $815. The national average of a computed tomography scan is $832. And the national average of a cardiac catheterization without coronary angiogram is $3755.[8] Also, what is the cost of making the wrong diagnosis? It is more than the financial cost of the procedure; it is the cost of inappropriate treatment, delayed diagnosis, and potential harm to the patient.[7]

The final argument to be presented here is the often said "well, we cannot do a RHC on everyone." This statement is not a guideline document determinant; it is a multifactorial societal, economic, and socioeconomic issue. Guidelines should influence the investment in infrastructure and mechanisms, such that the availability for an RHC universally should closely match its need.

The research question presented by Jansen et al. is reliant on current guidelines as a best practice diagnostic evaluation. The authors are to be commended on their study aimed to improve care in their country's community hospitals. They have proven that PH diagnosis needs to be comanaged with expert centers and that much of what is seen is Group 2 disease. Note, their data reflect common clinical practice patterns where hemodynamic measurements are performed *at the end* of the diagnostic workup (if at all). A straight line can be drawn from the myriad PH guidelines, including those participated in by the authors of this editorial, advocating hemodynamics as the last test, and *only* if pulmonary arterial hypertension is suspected. Herewith

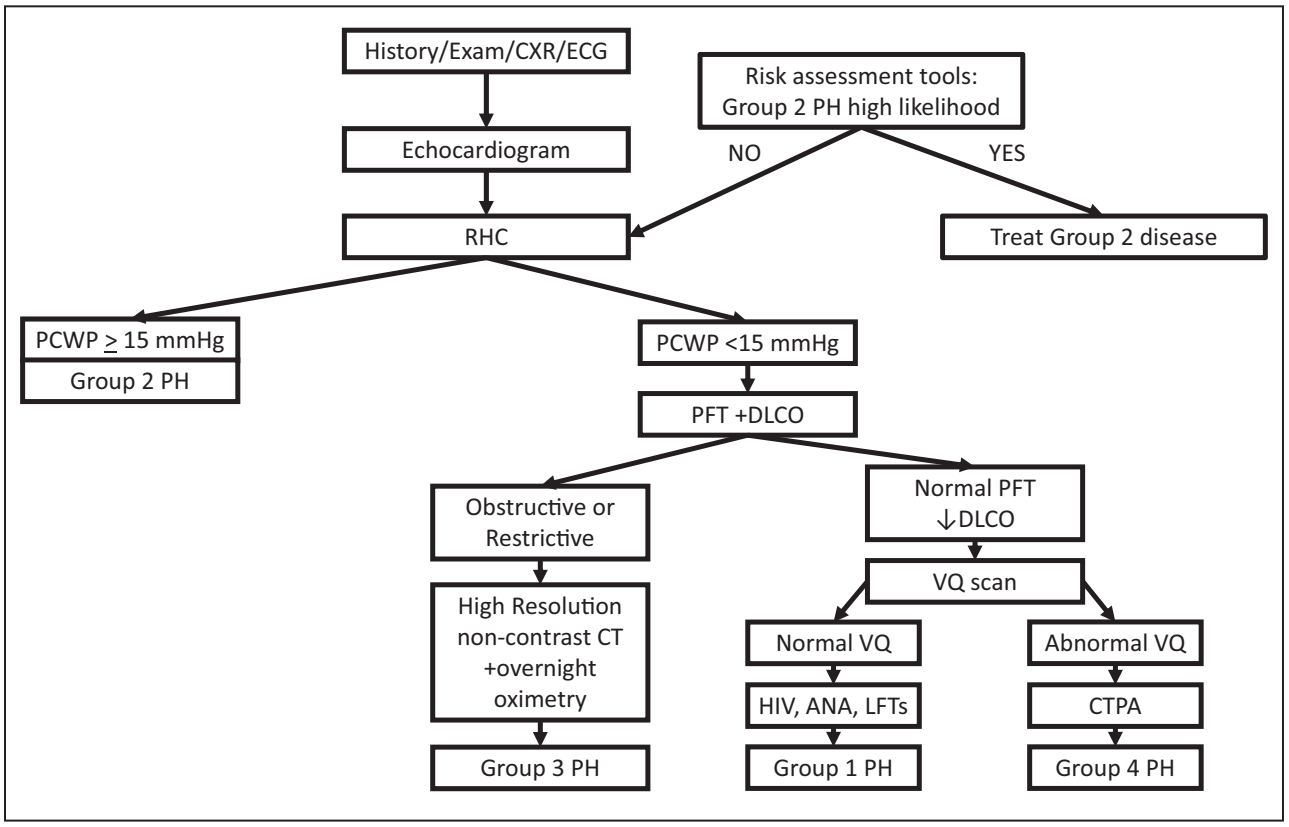

Figure. The Utah-George Washington Diagnostic Algorithm for pulmonary hypertension.
ANA indicates antinuclear antibody; CT, computed tomography; CTPA, CT pulmonary angiogram; CXR, chest x-ray; DLCO, diffusion capacity; LFTs, liver function tests; PCWP, pulmonary capillary wedge pressure; PFT, pulmonary function test; PH, pulmonary hypertension; and VQ, ventilation-perfusion scan.

are a series of statements from *evidence-based* guideline documents recommending diagnostic measures:

1. 2004 Screening, Early Detection, and Diagnosis of Pulmonary Arterial Hypertension: American College of Chest Physicians Evidence-Based Clinical Practice Guidelines: *Right-heart catheterization not only provides important indices of disease severity, but it also enhances the diagnostic process by excluding other etiologies such as intracardiac or extracardiac shunts and left-heart disease and provides an assessment of the degree of right-heart dysfunction through measurement of RAP [right atrium pressure] and cardiac output*.[9]
2. 2009 American College of Cardiology Foundation/American Heart Association Expert Consensus Document on Pulmonary Hypertension: *Some patients initially suspected of having PAH [pulmonary arterial hypertension] will not require catheterization, having had an alternative diagnosis established by noninvasive testing*.[2]
3. 2015 European Society of Cardiology/European Respiratory Society Guidelines for the diagnosis and treatment of pulmonary hypertension: *Cardiac catheterization should be performed after the completion of other investigations so that it can answer specific questions that may arise from these investigations and avoid an unnecessary procedure where an alternative diagnosis is revealed*.[7,10]

The Netherlands consortium reaffirmed the inability to complete our current diagnostic PH workup as it is currently recommended. Further discussion about the diagnostic algorithm is especially important as the updated European Society of Cardiology/European Respiratory Society guidelines will be published shortly. Contrary to the 2004 CHEST guidelines' own diagnostic algorithm, the authors make a compelling case for the role of RHC, not only in evaluating for pulmonary arterial hypertension, but also to assess for left sided heart disease. However, the prevalence of left sided heart disease as the predominant cause of PH is the very reason that the 2009 American College of Cardiology Foundation/American Heart Association document argues against doing an RHC upfront. Finally, the comment from the 2015 European Society of Cardiology/European Respiratory Society guidelines directly contributes to the 43% of patients in the study from Jansen et al. who had PH but never had RHC.[6]

It seems time to change our thinking. The proposed alternative diagnostic algorithm, titled, "The Utah-George Washington Diagnostic Algorithm," is based on our collective clinical experience (Figure). Although similarly no more supported by evidence than any of the other algorithms, the Utah-George Washington Diagnostic Algorithm brings the RHC to the forefront and separates patients based on proven diagnostic findings rather than performance of multiple tests to rule out everything before the RHC. There will be clinical circumstances where the likelihood of Group 2 PH is so high that upfront treatment without further workup is warranted.[11,12] However, oncologists insist on a definitive diagnosis with a biopsy before treatment is started because of the disparate treatment options and variants within cancer. It would be uncommon for a lung cancer guideline document to err *away from* sampling a malignant appearing mass to "*avoid an unnecessary procedure where an alternative diagnosis is revealed.*" We encourage those working in the PH field to rethink their approach to diagnosis and rather than deferring the definitive hemodynamic assessment to the end of the workup, they should make the right choice from the very beginning.

ARTICLE INFORMATION

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Disclosures

None.

REFERENCES

1. Rubin LJ. Primary pulmonary hypertension. *Chest*. 1993;104:236–250. doi: 10.1378/chest.104.1.236
2. McLaughlin VV, Archer SL, Badesch DB, Barst RJ, Farber HW, Lindner JR, Mathier MA, McGoon MD, Park MH, Rosenson RS, et al. ACCF/AHA 2009 expert consensus document on pulmonary hypertension: a report of the American College of Cardiology Foundation Task Force on Expert Consensus Documents and the American Heart Association: developed in collaboration with the American College of Chest Physicians, American Thoracic Society, Inc., and the Pulmonary Hypertension Association. *Circulation*. 2009;119:2250–2294. doi: 10.1161/CIRCULATIONAHA.109.192230
3. Ryan JT, de Jesus Perez VA, Ryan JJ. Health disparities in pulmonary arterial hypertension and the impact of the COVID-19 pandemic. *Adv Pulm Hypertens*. 2021;20:6–15. doi: 10.21693/1933-088X-20.1.6
4. Ryan JJ, Melendres-Groves L, Zamanian RT, Oudiz RJ, Chakinala M, Rosenzweig EB, Gomberg-Maitland M. Care of patients with pulmonary arterial hypertension during the coronavirus (COVID-19) pandemic. *Pulm Circ*. 2020;10:2045894020920153. doi: 10.1177/2045894020920153
5. Ryan JJ, Rich JD, Thiruvoipati T, Swamy R, Kim GH, Rich S. Current practice for determining pulmonary capillary wedge pressure predisposes to serious errors in the classification of patients with pulmonary hypertension. *Am Heart J*. 2012;163:589–594. doi: 10.1016/j.ahj.2012.01.024
6. Jansen SMA, Huis in 't Veld AE, Tolen PHCG, Jacobs W, MHM W, Grotjohan HP, Waskowsky M, van der Maten J, van der Weerdt A, Hoekstra R, et al. Clinical characteristics of patients undergoing right heart catheterizations in community hospitals. *J Am Heart Assoc*. 2022;11:e025143. doi: 10.1161/JAHA.121.025143
7. Deano RC, Glassner-Kolmin C, Rubenfire M, Frost A, Visovatti S, McLaughlin VV, Gomberg-Maitland M. Referral of patients with pulmonary hypertension diagnoses to tertiary pulmonary hypertension centers:

the multicenter RePHerral study. *JAMA Intern Med*. 2013;173:887–893. doi: 10.1001/jamainternmed.2013.319
8. Available at: https://www.mdsave.com/. Accessed May 30, 2022.
9. McGoon M, Gutterman D, Steen V, Barst R, McCrory DC, Fortin TA, Loyd JE; American College of Chest Physicians. Screening, early detection, and diagnosis of pulmonary arterial hypertension: ACCP evidence-based clinical practice guidelines. *Chest*. 2004;126:14S–34S. doi: 10.1378/chest.126.1_suppl.14S
10. Galie N, Humbert M, Vachiery JL, Gibbs S, Lang I, Torbicki A, Simonneau G, Peacock A, Noordegraaf AV, Beghetti M, et al. 2015 ESC/ERS guidelines for the diagnosis and treatment of pulmonary hypertension: the Joint Task Force for the Diagnosis and Treatment of Pulmonary Hypertension of the European Society of Cardiology (ESC) and the European Respiratory Society (ERS): endorsed by: association for European Paediatric and Congenital Cardiology (AEPC), International Society for Heart and Lung Transplantation (ISHLT). *Eur Respir J*. 2015;46:903–975. doi: 10.1183/13993003.01032-2015
11. Jansen SMA, Huis In 't Veld AE, Jacobs W, Grotjohan HP, Waskowsky M, van der Maten J, van der Weerdt A, Hoekstra R, Overbeek MJ, Mollema SA, et al. Noninvasive prediction of elevated wedge pressure in pulmonary hypertension patients without clear signs of left-sided heart disease: external validation of the OPTICS risk score. *J Am Heart Assoc*. 2020;9:e015992. doi: 10.1161/JAHA.119.015992
12. Thenappan T, Shah SJ, Gomberg-Maitland M, Collander B, Vallakati A, Shroff P, Rich S. Clinical characteristics of pulmonary hypertension in patients with heart failure and preserved ejection fraction. *Circ Heart Fail*. 2011;4:257–265. doi: 10.1161/CIRCHEARTFAILURE.110.958801