

Quality & Outcomes Research in Pulmonary Arterial Hypertension

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**University of Utah, Division of Cardiovascular Medicine.
American Heart Association Scientific Sessions 2014, Chicago, IL**

Emerging Researchers from 3CPR Council:

**How to Build a Road Map for Your Career in Cardiopulmonary, Critical Care,
Perioperative and Resuscitation Research**

#AHA2014

@drjohnjryan

November 15th 2014

@American_Heart

Outcomes Research

- “the study of the end results of health services that takes patients’ experiences, preferences, and values into account — is intended to provide scientific evidence relating to decisions made by all who participate in health care.”

Randomized Controlled Trials in PAH

- Instrumental in identifying the vast majority of FDA-approved therapies.
- Heterogeneous range of PAH substrates accounts for inconsistent rates of clinical benefit.
- RCTs have been successful at providing outcome data despite the (relatively) low prevalence of PAH.

#AHA2014

Monday Nov 17, 2014 9:00 AM - 5:00 PM

Poster Board Number: 5201

PAH Registries: 1981-present

| PH Registry | Date of Enrollment | Sample Size |
|---------------------|--------------------|-------------|
| NIH registry (3) | 1981–1988 | 194 |
| PH connection (6) | 1982–2006 | 578 |
| French registry (7) | 2002–2003 | 674 |

| | | |
|------------------------------------|-----------|------|
| REVEAL Registry (5) | 2006–2007 | 2525 |
| PAH registry in China (8) | 1999–2004 | 72 |
| PAH registry in UK and Ireland (1) | 2001–2009 | 482 |

Pulmonary Hypertension Registry of the UK & Ireland

- Aim: To create a registry of incident PAH.
- 482 patients:
 - 93% idiopathic, 5% heritable, &
- REVEAL risk score, and PH Connors survival equation accurately predicted survival
 - 1-year survival: 93%.
 - 5-year survival: 61%.



Yi Ling:

Then - Pulmonary vascular research fellow at SPVU.

Now - recently completed MD.

A study of PH misdiagnosis

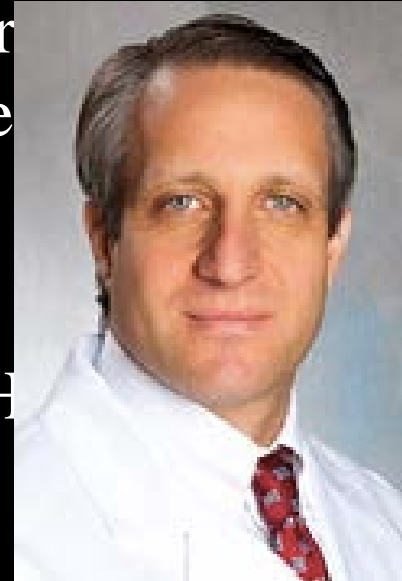
- Aim: Determine the accuracy of PH diagnoses in patients referred to PH centers.
- 140 patients:
 - mean age of 56 years.
 - 86 (61%) were WHO FC III or IV.
- 33% had received a misdiagnosis.
- 57% of the prescriptions contrary to published guidelines.



*Rod Deano:
Then- Internal
Medicine
Resident at
UChicago.
Now- Cardiology
fellow NYP.*

The heterogeneity of clinical practice patterns PAH experts

- Aim: to characterize the clinical management strategies used by PAH experts in PAH diagnosis, drug therapy, and the clinical assessment.
- Marked disagreement in the role of RHC in PAH management beyond diagnosis.
- Considerable agreement in favor of echocardiography for the routine monitoring of right ventricular function.

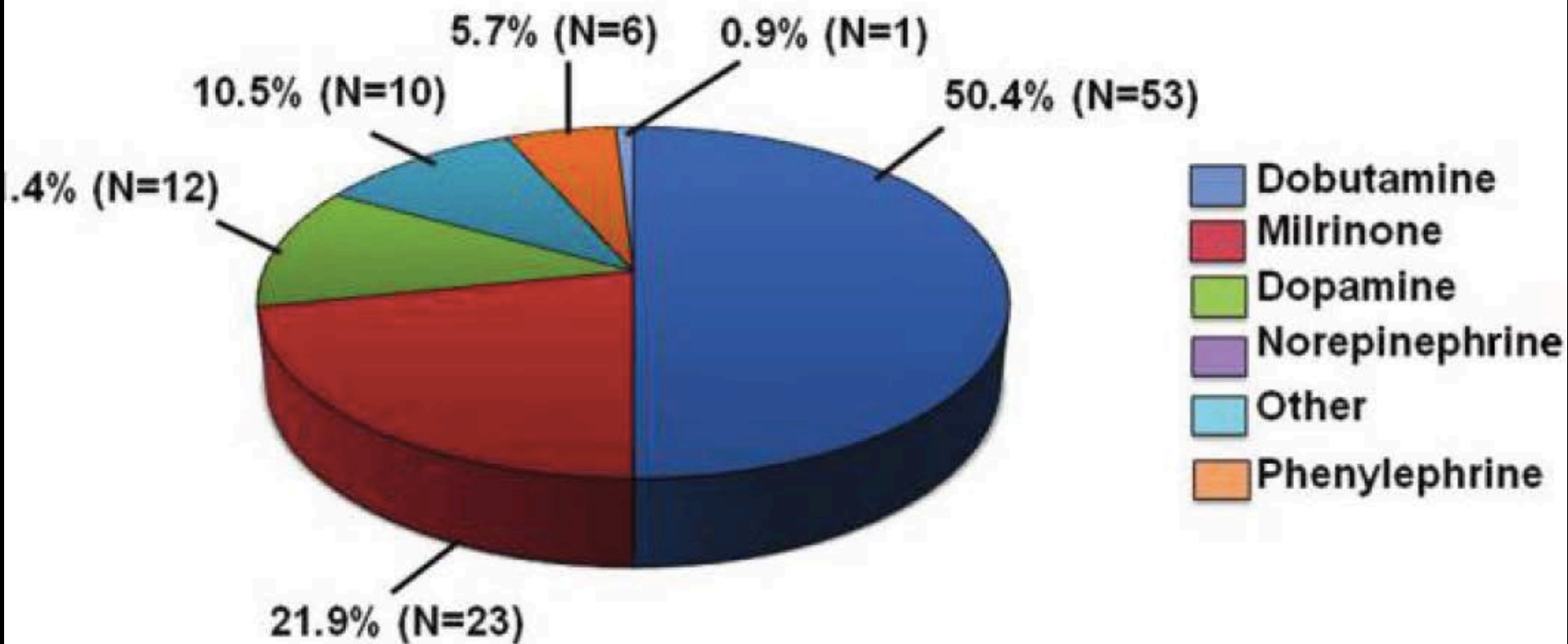


*Brad Maron:
Then - BWH.
Now - BWH.*

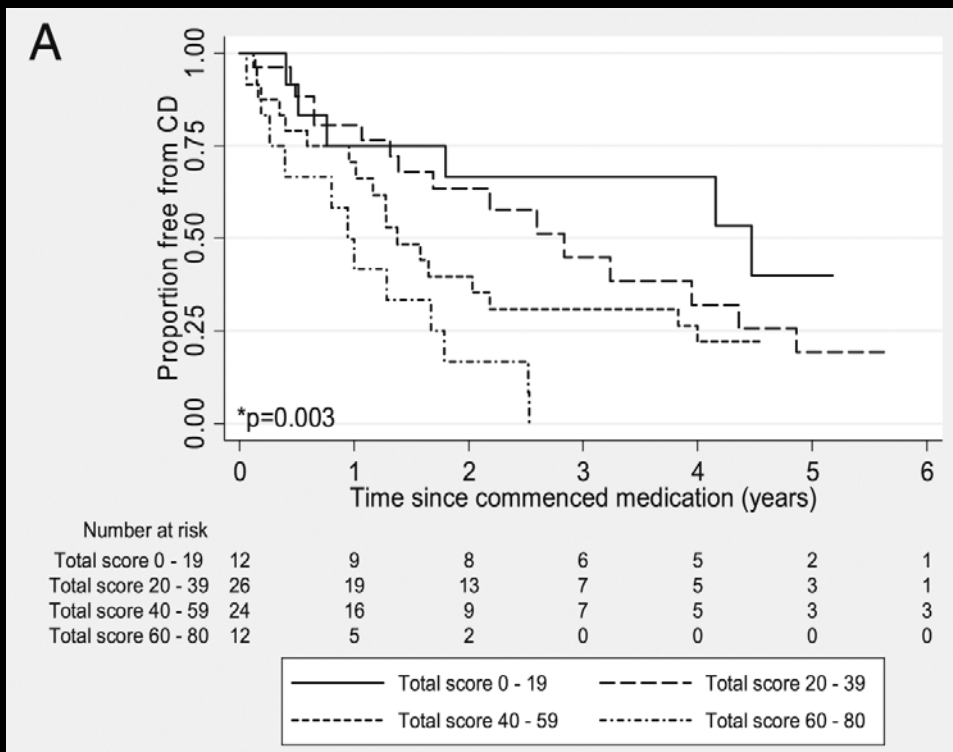
Role of Left heart catheterization in PAH

| LHC survey question | Response, N (%) |
|--|--------------------|
| LHC is not a part of the cardiopulmonary hemodynamic assessment in my practice | 58 (55.2) |
| LHC is always performed unless contraindicated | 30 (28.5) |
| LHC is performed only if the transpulmonary gradient is ≥ 10 mmHg | 6 (5.7) |
| LHC is performed only if the pulmonary capillary wedge pressure is < 15 mmHg | 5 (4.7) |

Preferred Inotrope in RVF from PAH



Cambridge Pulmonary Hypertension Outcome Review (CAMPHOR)



**Colm McCabe:
 Then – Research
 Fellow, PVDU.**

**Now -
 Consultant
 Respiratory
 Physician,**

Reappraisal of the NIH risk stratification equation

- In 576 patients with PAH referred during 2007.
- The observed 1-, 3- and 5-yr survival were significantly higher than the predicted survival.
- A new survival prediction equation was developed
 - $P(t) = e^{-A(x,y,z)t}$.
 - where P(t) is probability of survival.
 - t the time interval in years.

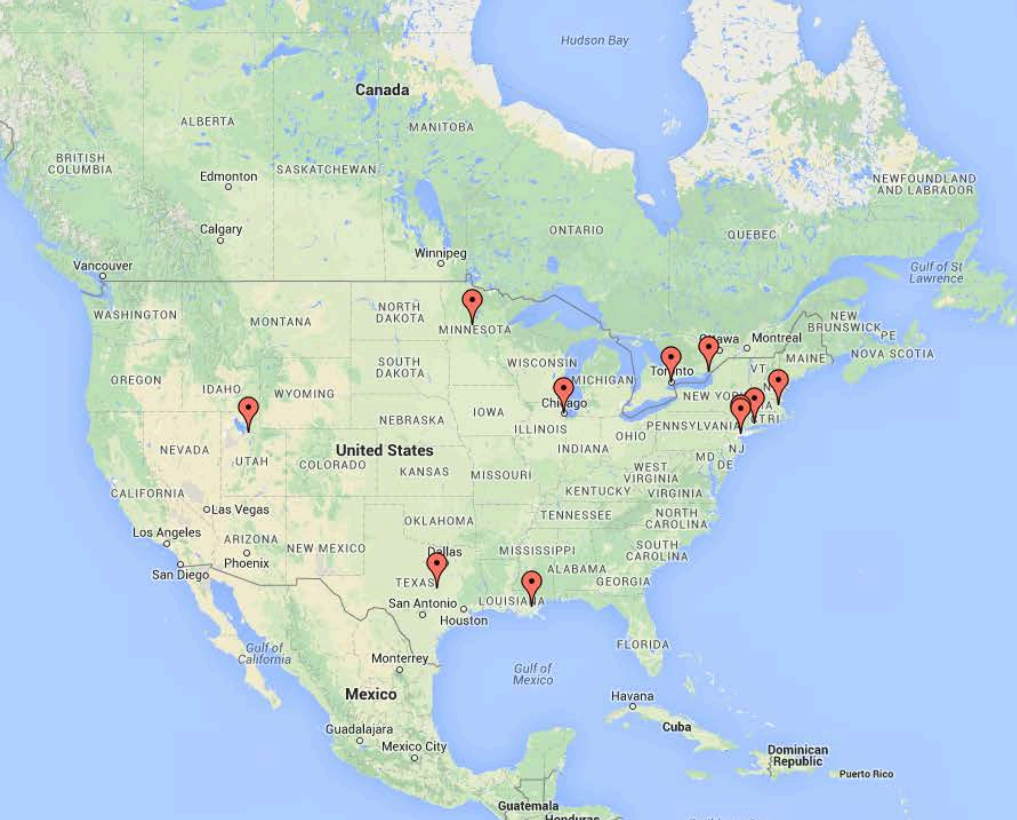


*Thenappan
Thenappan:
Then - PGY-4
Research fellow
at UChicago.
Now – Asst Prof,
UMinnesota.*

Summary

- The field of PAH has heavily depended on registries & RCTs which has given insight into this rare disease.
- Registries & Surveys offer insight to clinical practice.
- Young investigators and early career researchers have led this field of research in PAH.

*3CPR Early Career Networking Lunch,
Tuesday, November 18th 12:00pm - 1:30 pm:
Hyatt Regency McCormick Place, Grant Park AB.*



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Queen's University
Stephen Archer, MD

University of Utah
Nathan Hatton, MD
Jess Harris, MD

University of Minnesota
Thenappan Thenappan, MD

University of Kent
Ghazwan Butrous, MD

Yale University
Harlan Krumholz, MD
Emily Bucholz, MS

Harvard Medical School
Bradley Maron, MD

St Luke's Roosevelt
Saurav Chatterjee, MD

Northwestern University
Jonathan Rich, MD **#AHA2014**
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