Diagnostic Dilemmas in Pulmonary Hypertension

Michael D. McGoon

Professor of Medicine

Consultant, Cardiovascular Diseases

Mayo Clinic

Rochester, MN

mmcgoon@mayo.edu

Dilemmas

- How accurate is the echocardiogram for assessing pulmonary hypertension?
- Is measurement of exercise hemodynamics useful?
- How should borderline pulmonary hypertension be approached?
- Which outcome predictor is best?
- How should patients be followed?

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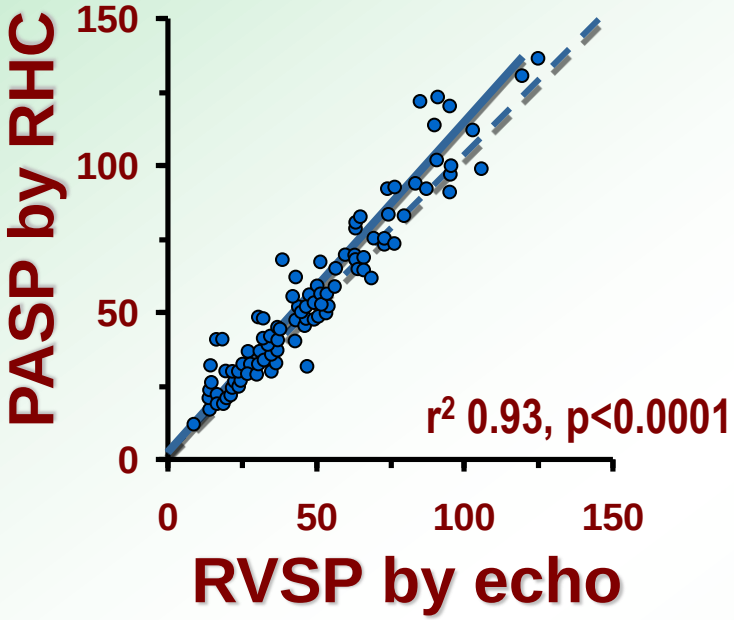
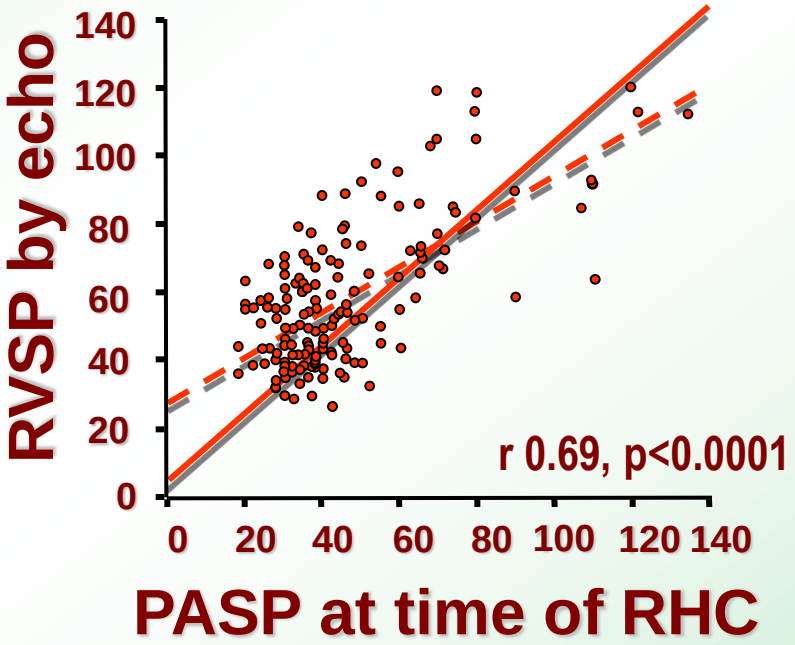
How Accurate is the Echocardiogram?



Simultaneous RHC and Doppler
Currie et al: JACC 6:750, 1985



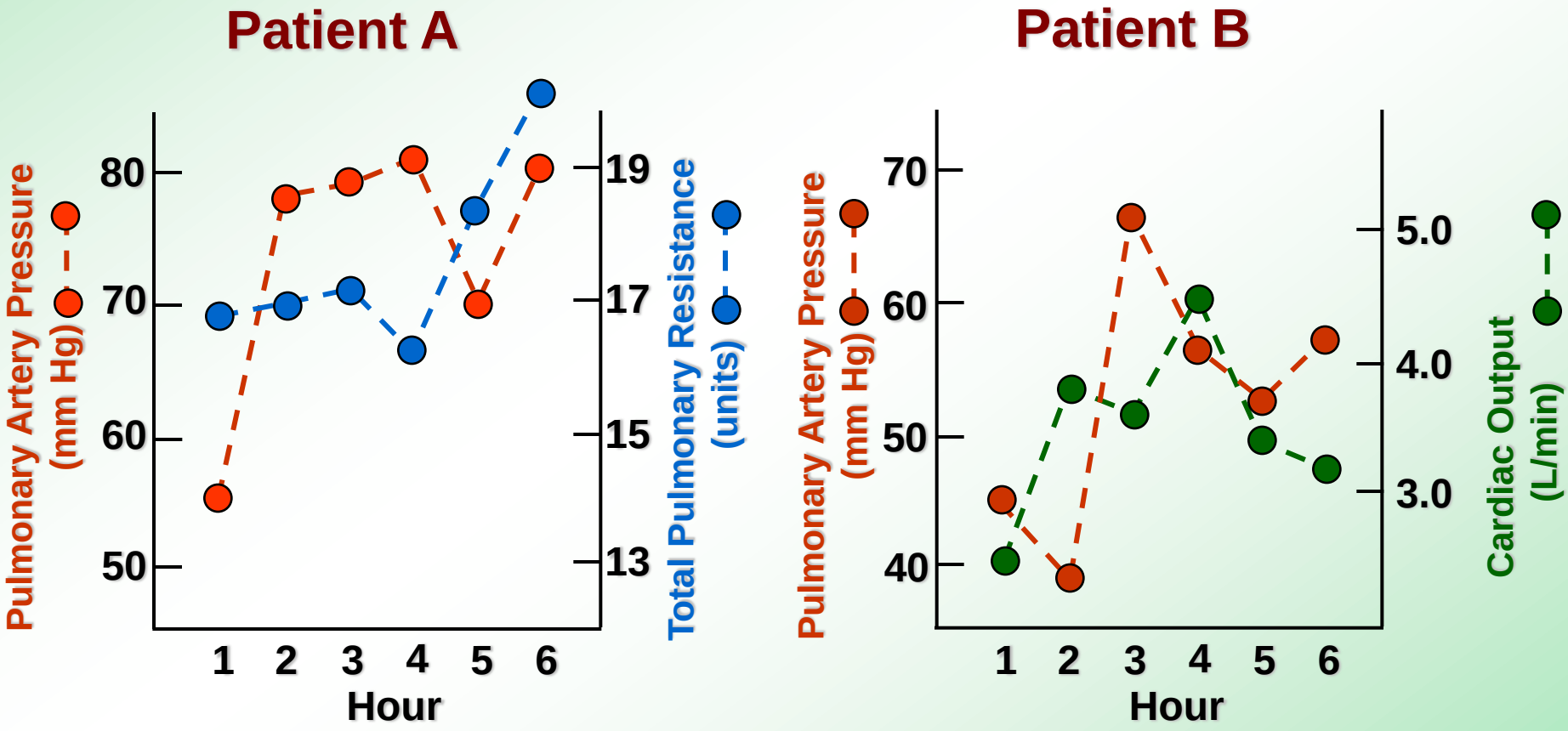
RHC and Doppler within 72 hours
Arcasoy et al; AJRCCM 2003; 167:735-40

How Accurate is the Echocardiogram?

Reasons for failure of echo-cath RVSP correlation

- Underestimating TR velocity – not getting peak signal, poor alignment
- Overestimating TR velocity – taking overgained shaggy signals
- Failing to adequately assess right atrial pressure
- Cath inaccuracies
- Changing conditions – changing pressures

Spontaneous Variation in Pulmonary Arterial Hemodynamics Over Time in Two Representative Patients



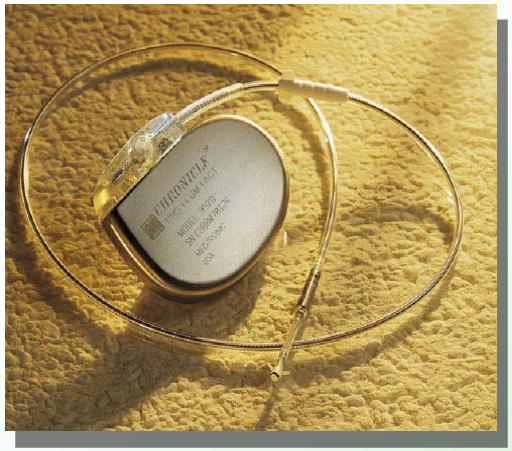
Rich et al. AJC 1985 55(1): 159-163.

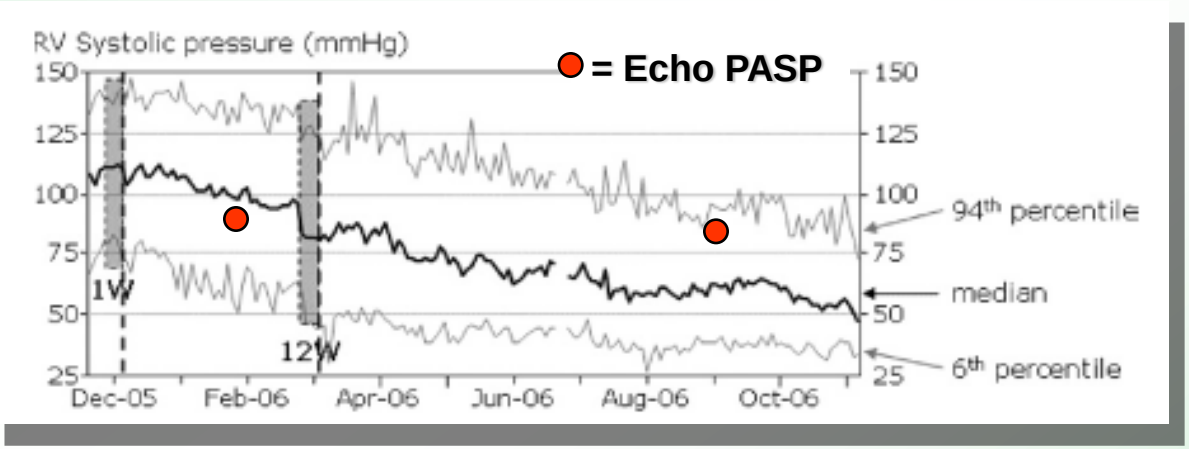
How Accurate is the Echocardiogram?

Implantable Hemodynamic Monitor: Chronicle®

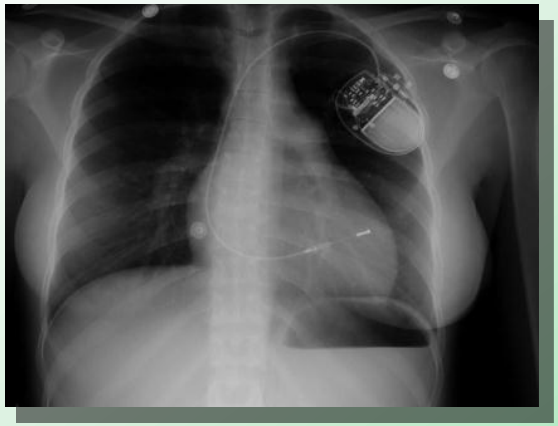
RV pressure sensor measures:

- Systolic and diastolic pressure
- Estimates PA diastolic pressure
- RV dp/dt
- Heart rate, temperature, activity





Based on Frantz RP et al. J Heart Lung Transplant 2008



How Accurate is the Echocardiogram?

Demographics

Height: 159.9 cm **Weight:** 69.0 kg **BSA:** 1.72 m² **BMI:** 26.99
Procedure Start Time: 07/22/2009 09:01 AM **Location:** GO 6S
Referring Provider: McGoon, Michael D **Indication for Study:** pulmonary hypertension;

Procedure

Type: Adult TTE
Components: 2–D, Color Flow Doppler, Doppler, M–Mode, TDI (Tissue Doppler Imaging)

Referral Diagnosis

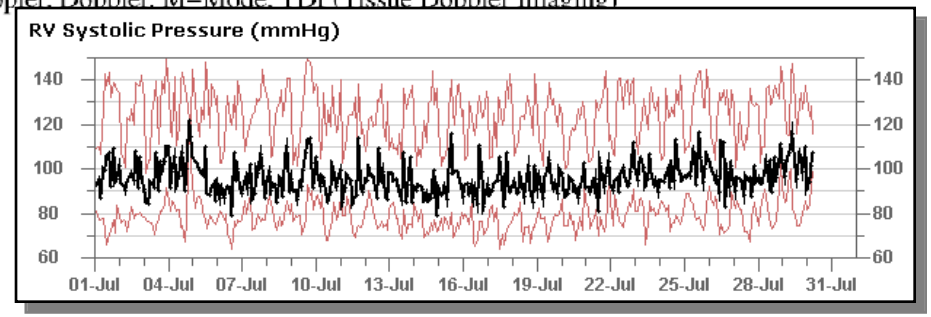
Pulmonary hypertension.
Mitral stenosis.

Hemodynamics

Heart Rate:
Blood Pressure:
ECG:

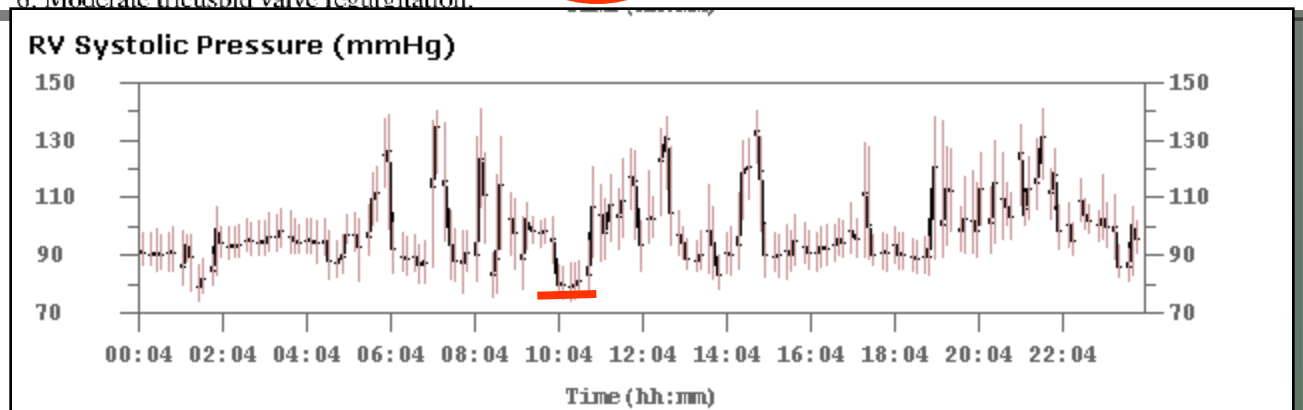
Media Details

Server #clinical clips = 88



Final Impressions

1. Normal left ventricular chamber size.
2. Calculated left ventricular ejection fraction; 71 %.
3. Severe left atrial enlargement.
4. Mitral valve diastolic mean Doppler gradient: 7 mmHg (heart rate 54 BPM). Down from 8mmHg.
5. Estimated right ventricular systolic pressure; 75 mmHg. This is decreased from 114mmHg exam of 02/02/2009.
6. Moderate tricuspid valve regurgitation.



McGoon – unpublished data

Reliability of Right Heart Catheterization

- Accurate at one point of time under a unique set of circumstances
  - Hemodynamics fluctuate over short periods of time
  - Hemodynamics fluctuate under varying circumstances
- Instantaneous PASP in PH is like fasting plasma glucose in diabetes
  - Accurate
  - May not be representative
- Representative measures (Hgb A1c in diabetes)
  - Daily median pulmonary arterial pressure – not available
  - Symptoms – Functional class, exercise capacity (6MWD)
  - RV function – cardiac MR or CT, BNP, TAPSE, RV strain

“Never Treat a Single Number!”

Echo and RHC – Complementary Info

Echo

- Other substrates
- Estimated RVSP, PAm, PAd, SV/PP, RV dP/dt, PVR
- RV Function
  - Visual
  - RV-IMP
  - TDI
    - TAPSE
    - Strain, Strain Rate
    - Isovolumic Accel

Screening & Follow-up

RHC

- Full hemodynamics
  - PCWP
  - Pulmonary Blood Flow
  - RAPm
- Vasodilators Response
- Oxygen saturations
  - Shunt evaluation

Definitive Diagnosis & Confirmation

Dilemmas

- How accurate is the echocardiogram for assessing pulmonary hypertension?
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Usefulness of Exercise Hemodynamics?

- Normal exercise mPAP is 30–45 mm Hg
- Clinical implication of higher values is uncertain

Usefulness of Exercise Hemodynamics?

- Depends on circumstances
  - Screening of high risk patients
  - Diagnosis of symptoms

Usefulness of Exercise Hemodynamics? Screening

- Early detection of PH in patients with:
  - Connective tissue disease?
  - BMPR2 mutation or family history?
- 2003: 3rd World Symposium on PH definition of PH (Venice)
  - mPAP >25 mmHg at rest
  - mPAP >30 mmHg with exercise
- 2008: 4th World Symposium on PH definition of PH (Dana Point, CA)
  - mPAP >30 not supported for screening

Usefulness of Exercise Hemodynamics? Problems with Screening

- Echocardiogram
  - Potentially inaccurate
  - Nonstandardized techniques
  - Does not distinguish possible exercise-related increase in pulmonary venous pressure due to left heart factors (diastolic dysfunction)
- Right Heart Catheterization
  - Not suitable for screening
  - Normal cut-off values uncertain in subpopulations (age, gender, fitness)
  - Implications of “abnormal” findings not known

Usefulness of Exercise Hemodynamics? Diagnosis of Source of Symptoms

- May be useful in patient with symptoms causing exercise intolerance without other explanation after full evaluation for:
  - Left ventricular failure
  - Myocardial ischemia
  - Exercise induced reversible airways disease
  - Arrhythmia/Chronotropic incompetence
  - Anemia
  - Deconditioning (cardiopulmonary exercise test)
- Requires confirmation with exercise RHC

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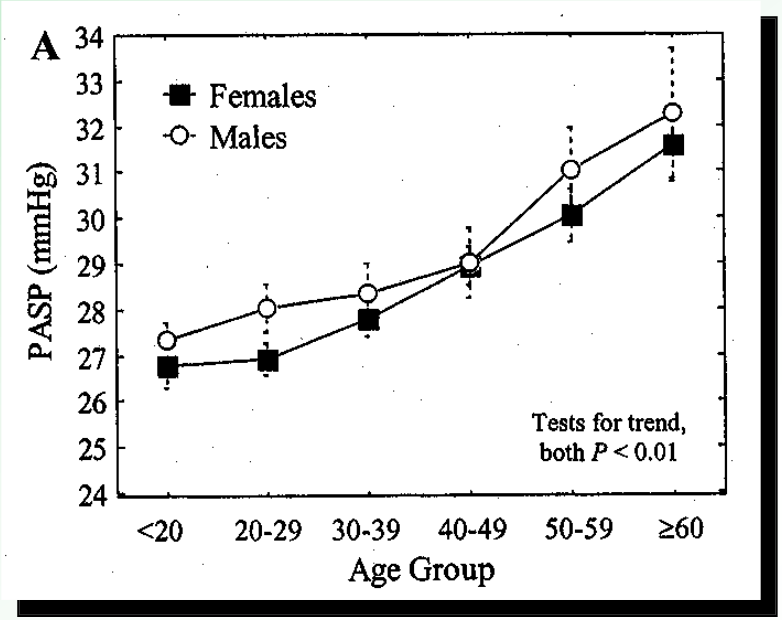
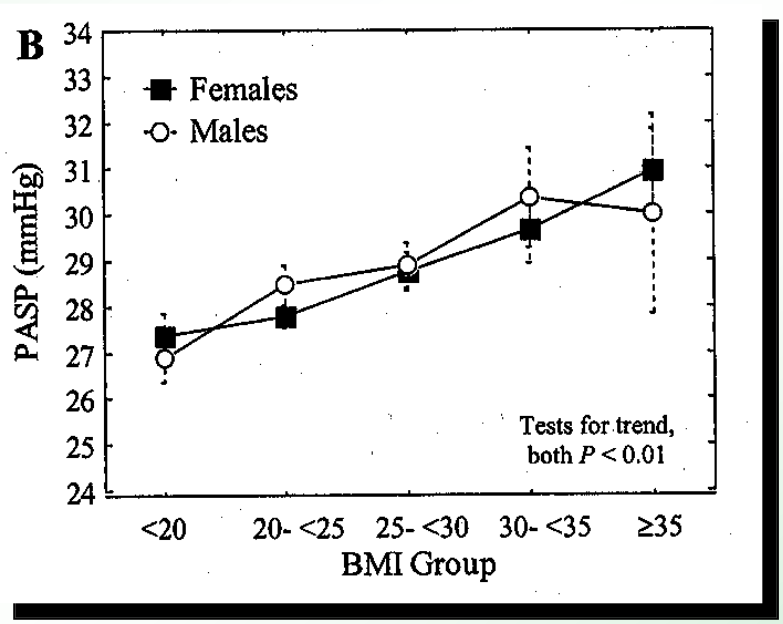
How to Approach Borderline PH?

- 62 year-old man with dyspnea on exertion
- Echocardiogram: RVSP 43 mmHg

How to Approach Borderline PH?

Cut-off for Normal is Not the Same for Everyone





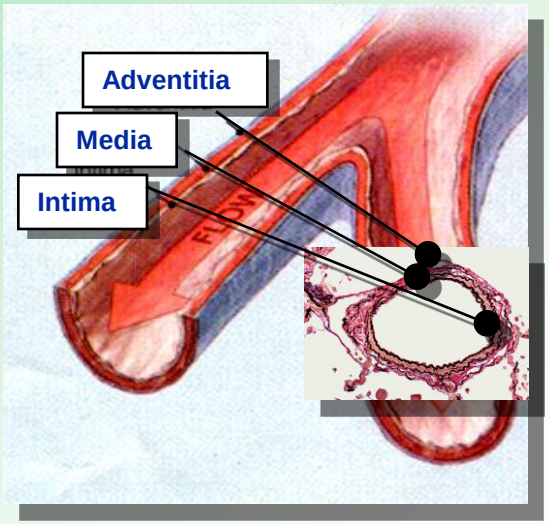
McQuillan et al: Circulation 2001;104:2792-2802

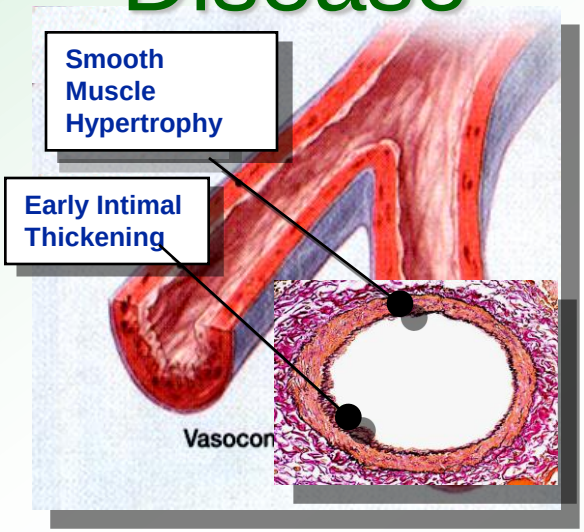
***MGH Cardiac US study:** 3790 normal TTE's with measured PASP*

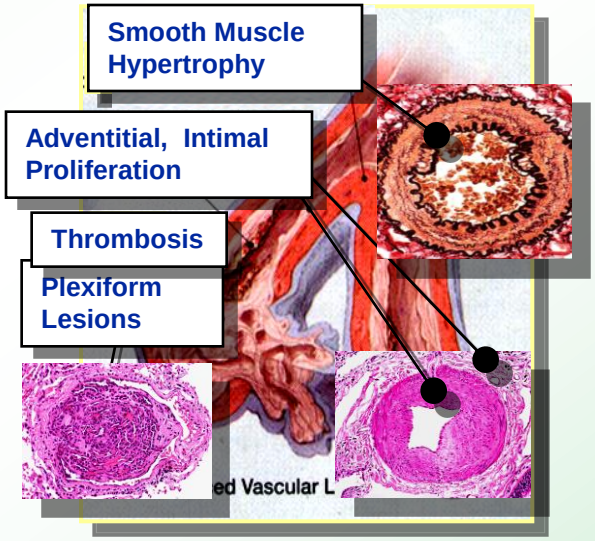
- **2,472 normal TTE's:**
  - **Mean PASP 33.1±7.7 mm Hg, 60% >30 mm Hg**
  - **People over 50 yo: 22% >40 mm Hg**
  - **People with BMI >30 Kg/m2: 20% >40 mm Hg**

Milani et al (Ochsner), ACC 2003

Progression of Pulmonary Vascular Disease







PVR (<3 U)	↑	↑
mPAP (<25 mmHg)	↑	↓
RV	↑stress, compensation, enlargement	failure
CO (>5 L)	maintained	↓

Implications of Borderline/Mild PH

- Understand the limitations of the measurement
- Determine whether there is another cause of symptoms
- Consider strength of corroborating information and risk profile of the patient
- May be a sign of EITHER early disease or late decompensated disease
- May be a sign of other issues: age, systemic hypertension, left heart disease, body mass
- Requires evaluation (if symptomatic) or follow-up (if asymptomatic)

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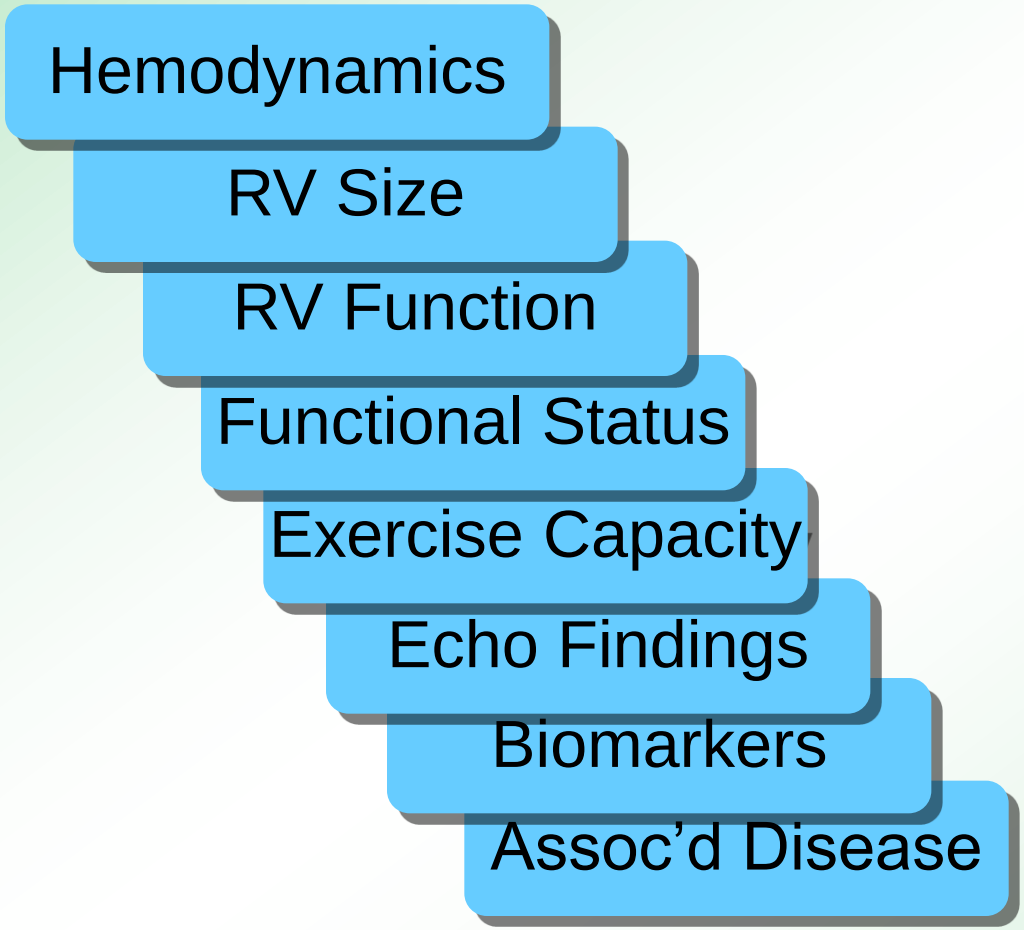
Predictors of Outcome



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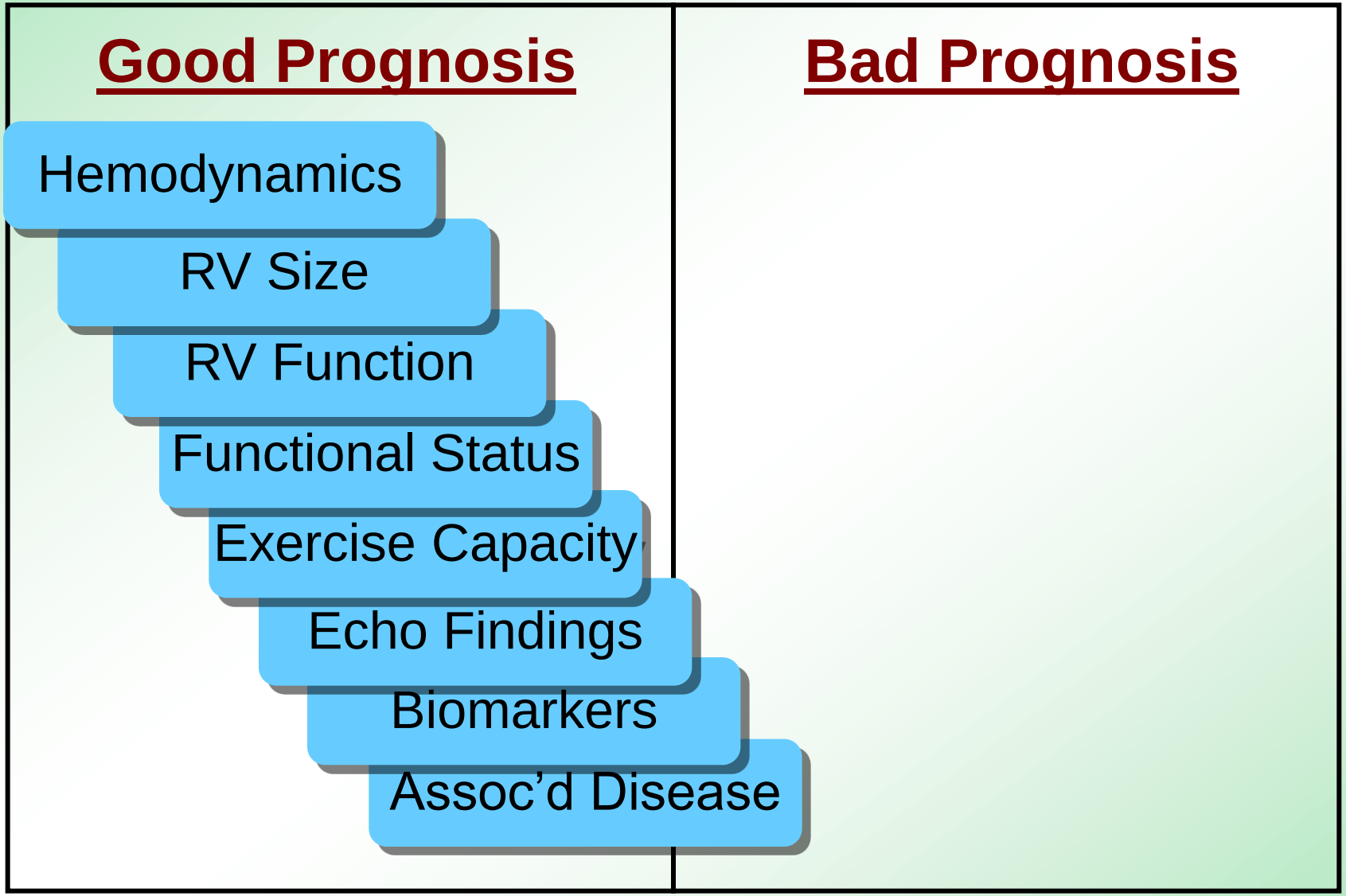
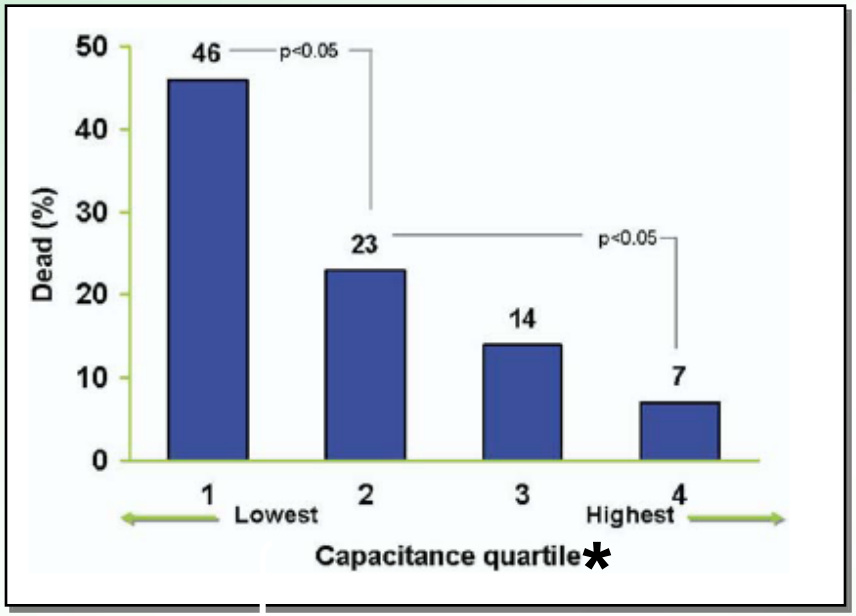
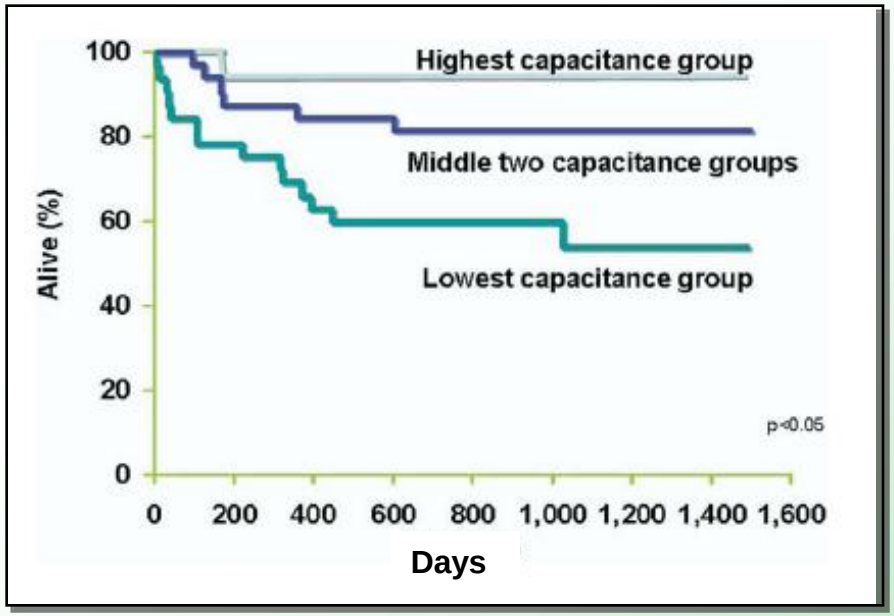


Predictors of Outcome

RV/PA Coupling

Pulmonary Vascular Capacitance ≈ SV/PP





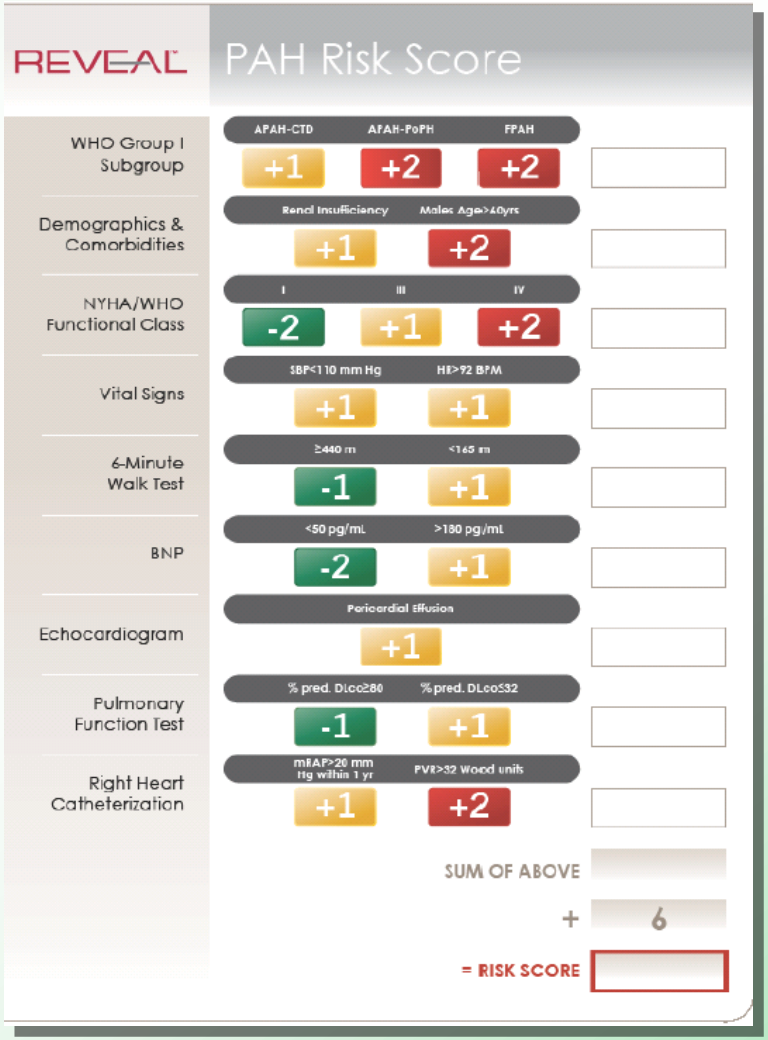
Lowest .38-.80
Next Lowest .81-1.24
Next Highest 1.25-2.08
Highest 2.09-3.59

Mahapatra et al. The Prognostic Value of Pulmonary Vascular Capacitance Determined by Doppler Echocardiography in Patients with Pulmonary Arterial Hypertension. JASE 2006;19(8):1045-50
Mahapatra et al. Relationship of Pulmonary Arterial Capacitance and Mortality in Idiopathic Pulmonary Arterial Hypertension. JACC 2006;47(4):799-803

Predictors of Outcome

Composite Models – REVEAL Registry

A risk calculator has been developed which uses commonly available information, but doesn't require that every variable is available in order to work

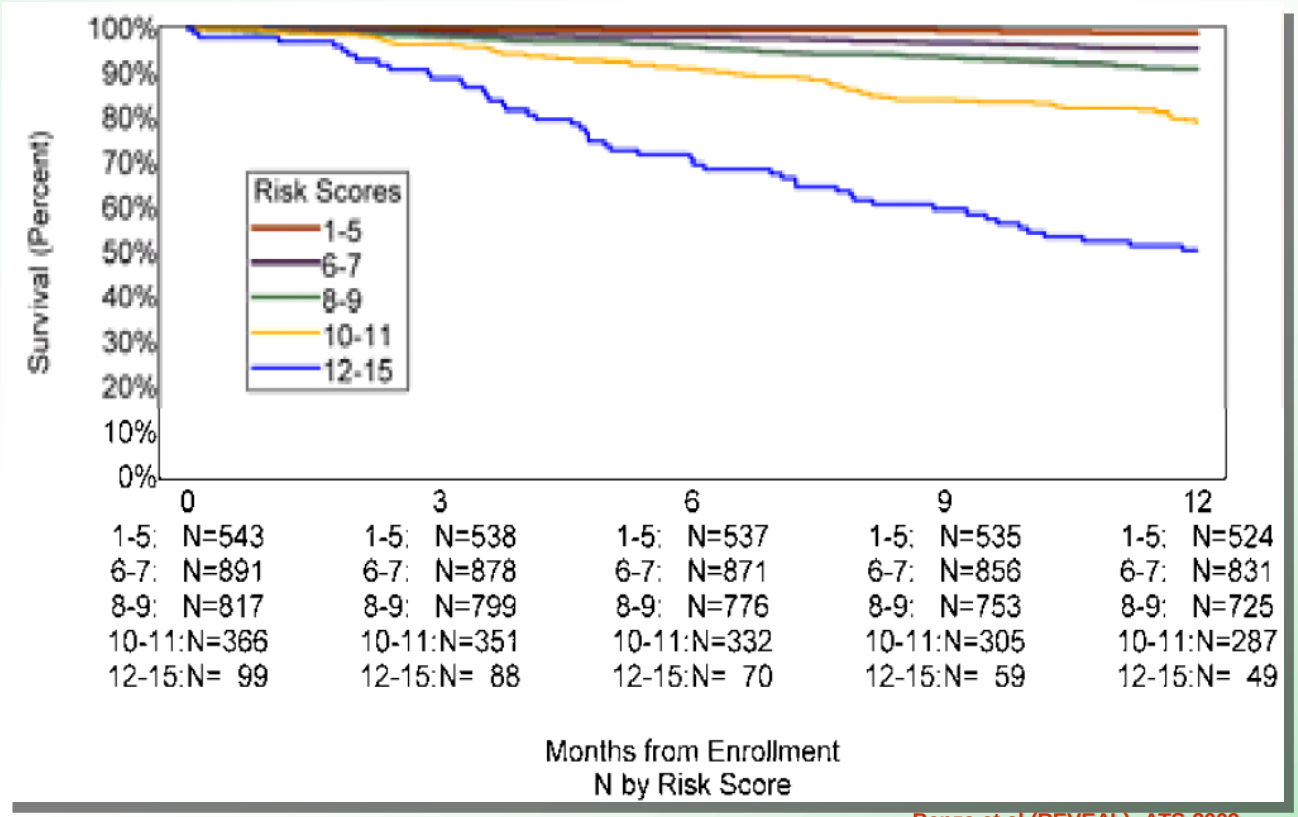


Benza et al (REVEAL): ATS 2009

Predictors of Outcome

Composite Models

Risk scores can distinguish different probabilities of survival for 1 year.



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Follow-up: When Should Treatment Be Changed?

- Treat-to-Target Strategy
  - Pre-specified outcome target
  - Based on improvement in risk
  - Targets
    - Systolic Blood Pressure >120 mmHg
    - MVO2 on Cardiopulmonary Exercise Test >14 ml/kg
    - 6MWD >360 meters