THE EARLY YEARS OF TREATMENT

- A few drugs available for an orphan disease
- Drugs chosen by functional class (III/IV)
- Treating early functional class was controversial
- 6MWT at 3-4 months was the primary endpoint of studies, not much known beyond that
PULMONARY ARTERIAL HYPERTENSION — A TREATMENT ALGORITHM

RCT FOR TREATMENT NAIVE PATIENTS

<table>
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<tr>
<th>Treatment</th>
<th>N</th>
<th>RR</th>
<th>p value</th>
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*AIR had a co-primary endpoint

RCT — BACKGROUND THERAPY ALLOWED

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PULMONARY ARTERIAL HYPERTENSION — A TREATMENT ALGORITHM

COMBINATION UPFRONT THERAPY — AMBITION

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PULMONARY ARTERIAL HYPERTENSION — A TREATMENT ALGORITHM

TRIPLE UP-FRONT COMBINATION RX


PULMONARY ARTERIAL HYPERTENSION — A TREATMENT ALGORITHM

HOW TO CHOOSE?

• Use the diagnostic process and testing to assess the patient’s short term risk of early mortality
• Use treatment guidelines to help determine initial choice(s)
• Follow a regular testing and visit schedule to both calculate ongoing risk and ensure the patient is meeting goals
• Change/add therapy if goals are not met or risk increases

PULMONARY ARTERIAL HYPERTENSION — A TREATMENT ALGORITHM

DETERMINANTS OF RISK

Anticoagulation ± Diuretics ± O2 ± Digoxin

Acute vasoreactivity testing (for Idiopathic PAH)

- Low/Intermediate risk: TPG or PAH-VRCP
- High risk: Combination including IV/SC Prostacyclin

Treated response?

- Low/Intermediate Risk: ERA or PDE5i or ERA + PDE5i
- High Risk: Combination including IV/SC Prostacyclin

Inadequate response:

Other options:
- Riociguat
- Oral PGI2 or IP antagonist
- Inhaled PGI2
- IV or SC PGI2

- 2-drugs up-front (with h/o)
- 3-drugs up-front (with h/o)

LONGITUDINAL PATIENT MONITORING: ACCF / AHA RECOMMENDATIONS

<table>
<thead>
<tr>
<th>Patient Evaluation</th>
<th>6-MWD</th>
<th>FC</th>
<th>BNP</th>
<th>ECHO</th>
<th>RHC</th>
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<td>Stable patient</td>
<td>Every 3-6 months</td>
<td>Every visit</td>
<td>Every 12 months</td>
<td>If clinical deterioration</td>
<td></td>
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<tr>
<td>Unstable patient</td>
<td>Every 1-3 months</td>
<td>Every visit</td>
<td>Center dependent</td>
<td>Every 6-12 months</td>
<td>Every 6-12 months or if deterioration</td>
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</table>


TREATMENT GOALS — CONSENSUS FROM 5™ WSPH

<table>
<thead>
<tr>
<th>6-MWD</th>
<th>CPET</th>
<th>FC</th>
<th>BNP</th>
<th>ECHO</th>
<th>Hemodynamics</th>
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</thead>
<tbody>
<tr>
<td>&lt; 380 – 440 meters</td>
<td>Peak VO2 &gt; 15 mL/min/kg</td>
<td>EtCO2 &lt; 45 l/min</td>
<td>Normal or near normal RV size and function</td>
<td>RAP &lt; 8 mm Hg</td>
<td>CI &gt; 2.5 - 3 L/min/m²</td>
</tr>
</tbody>
</table>

INTERVENTIONAL PROCEDURES: BALLOON ATRIAL SEPTOSTOMY

- Creation of an interatrial right-to-left shunt
- In order to:
  - Decompress right heart chambers
  - Increase LV preload
  - Increase CO
  - Improve systemic oxygen transport
  - Decrease sympathetic hyperactivity
- Considered a palliative or bridging procedure
- Patients refractory to medical therapy
- Patients awaiting lung transplantation


ISHLT GUIDELINES FOR LUNG TRANSPLANTATION FOR PAH

- Persistent FC III or IV despite maximal medical therapy
- Low (< 350 meters) or declining 6-MWD
- Failing even while on a parenteral prostacyclin analog
- CI < 2 L/min/m²
- RAP > 15 mm Hg

ISHLT = International Society for Heart Lung Transplantation

SUMMARY

- The treatment algorithm for pulmonary arterial hypertension has changed over the last few years to be more proactive rather than reactive in this progressive disease
- Regular follow up for PAH patients with assessment of the goals of treatment will allow revision or addition to the treatment strategy proactively
Drug Therapies for PAH

James R. Klinger, MD
Division of Pulmonary, Sleep and Critical Care Medicine
Rhode Island Hospital
Brown University
Providence, RI

Faculty Disclosures

- Research support/grants from Actelion, Bayer, Eiger, Gilead, Ikaria, Lung LLC, NIH-NHLBI, and United Therapeutics
- Consultant for Bayer and United Therapeutics

Pharmacologic Therapies for PAH
Mechanisms of Action of Therapies

**Endothelin Pathway**
- Endothelin-1
- Pre-proendothelin
- Endothelin receptor A
- Endothelin receptor B

**Nitric Oxide Pathway**
- Nitric Oxide
- Arginine
- Eicosanooids

**Prostacyclin Pathway**
- Prostacyclin (prostaglandin I₂)
- Prostanoid PDE5 Inhibitor

**S.G.**
- I.V. Infusion
- Treprostinil (Remodulin)
- Epoprostenol (Flolan, Veletri (Generic))

**I.V.**
- Treprostinil (Remodulin)

**Oral**
- Treprostinil (Orenitram)
- Sildenafil (Revatio)
- Tadalafil (Adcirca)
- Bosentan (Tracleer)
- Ambrisentan (Letairis)
- Macitentan (Opsumit)
- Riociguat (Adempas)

**Inhaled**
- Treprostinil (Tyvaso)
- Iloprost (Ventavis)

**S.Q.**
- Treprostinil (Remodulin)

**Goals of Therapy for PAH**

**Improve pulmonary hemodynamics?**
- Pulmonary artery pressure
- Cardiac output

**PAP/CO = PVR**

**Improve exercise capacity?**
- 6 min walking distance
- WHO functional class

**Prevent disease progression?**
- Survival
- Time without clinical worsening
Impact of Approved Therapies on mean PA Pressure

Epoprostenol
Bosentan
Treprostinil
Iloprost
Macitentan
Ambrisentan
Tadalafil
Sildenafil
Riociguat
Selexipag

Impact of Therapy on PA Pressure

Impact of Therapy on Exercise Capacity

Delay in Time to Clinical Worsening*

Defined As:
- Death
- Lung transplantation
- Hospitalization for worsening PAH
- Initiation of IV prostacyclin therapy
- Decline in functional capacity (6 MWT, WHO FC)
Impact of Clinical Worsening on Survival in PAH

How Do We Choose?

STEP 1

Updated Treatment Algorithm of Pulmonary Arterial Hypertension

Pharmacologic Therapy for Pulmonary Arterial Hypertension in Adults

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)

STEP 1
STEP 2
Choosing Initial Therapy

STEP 3
What if they don’t get better?

Sequential Combination Goal-Directed Therapy
Combination Therapy for PAH

• Are 2 Drugs (or 3 drugs) Better Than 1?

THE AMBITION STUDY

500 Treatment naïve patients with WHO Group 1 PAH randomized 1:1:2 to receive:
- Tadalafil 40 mg/day + ambrisentan placebo
- Ambrisentan 10 mg/day + tadalfil placebo
- Tadalafil 40 mg/day + Ambrisentan 10 mg/day

Primary outcome measure: Time to first clinical failure defined as:
- Death
- Lung transplantation
- Hospitalization for worsening PAH
- Initiation of intravenous prostanoid /in therapy
- 15% Decline in 6-MWD

Time to First Clinical Failure

N Engl J Med. 2015 Aug 27;373(9):834-44
Risk Stratification and in PAH

Choice of initial treatment is driven by functional class and mortality risk. Mild to moderate impairment of function (FC II, III) with low 1 year mortality risk can be treated with oral therapy. Advanced disease (FC IV or FC III + high risk for increased 1 year mortality) should be treated with parenteral prostacyclin therapy. Up front combination therapy may be more efficacious than use of a single agent and should be considered in patients with increased risk. Patients who do not improve should be given additional therapy.

Summary and Key Points
IMPLANTABLE HEMODYNAMIC MONITORING IN THE MGMT OF PULMONARY ARTERIAL HYPERTENSION

RITA JERNY MO
DIRECTOR, CENTER FOR ADVANCED CARDIAC THERAPIES
ST FRANCIS HOSPITAL ROSELYN, NY

PAH AND AMBULATORY HEMODYNAMIC MONITORING

- CAN BE USED FOR BASELINE AND PERIODIC AMBULATORY HEMODYNAMICS
- USED TO MONITOR AND GUIDE RESPONSE TO THERAPY
- ASSESS CHRONICITY AND EVOLVING SEVERITY OF DISEASE
- FOLLOW TRENDS

PROS OF HEMODYNAMIC MONITORING

- PROVIDE MORE COMPLETE HEMODYNAMIC MEASUREMENTS IN THE HOME ENVIRONMENT UNDER DAY TO DAY CONDITIONS
- CAN PREDICT IMPENDING DECOMPENSATION
- PREVENT HOSPITALIZATIONS
- MORE ACCURATE ASSESSMENT OF BORDERLINE PH PATIENTS OR EXERCISE INDUCED SYMPTOMS
• SAFETY OF DEVICE IN POTENTIALLY HYPERCOAGULABLE PATIENTS
• TREATMENT ALGORITHMS ARENT STANDARDIZED

MA, PA et al. Continuous right heart pressure measurements with an implantable monitor. JCF, 2002;8(2):63-70

• MEDICATION CARDIOME AND SFACT
• THERAPY GUIDELINES ISSUED BY MH and EMPLOYEES & MANUFACTURERS
• CARDIOMEM DEVICES AND CARDIOMEM MONITORING
• PROOF OF CONCEPT STUDY WITH CARDIOMEM AND MH


Raymond Benza, Priscilla Correa-jaque, Veronica Franco, Mark Doyle, Jason A White, Greg Olin, Robert Biederman
Challenges in Pulmonary Hypertension (Focus is on PAH)

- It is a fatal disease
- Delayed diagnosis / misdiagnosis
- Inadequate evaluation (e.g., lack of RHC, VQ scan, etc)
- Delayed referral to PH Center
- Poor response to therapy in some patients
- Management of critically ill PH patients
- Pregnancy in PH patients
- Adverse effects of medications
- Inconvenience of most potent therapies
- Access to medications (less concern in U.S.)
- Expense of medications

DISCLOSURES:

- Research Funding (Cedars-Sinai Medical Center):
  - Actelion, Bayer, United Therapeutics, Reata, Arena, Eiger
- Consulting / Advisory Boards:
  - Actelion, Bayer, United Therapeutics, Reata
- Speaker Honoraria*
  - Actelion, Bayer

*Honoraria paid to Cedars-Sinai Medical Center
CASE 1: 36 year-old woman with SLE and PAH

- Dyspnea x 3 months.
- Echo showed “PH” – started on CCB
- Echo 2 months later - “No PH. Can get pregnant.”
- Echo at 20 weeks pregnant (DOE) - “PH is back”
- Refused termination
- PH Center eval neg for other causes except SLE
- Mildly hypoxemic, placed on 2L O2
- Echo and RHC performed
PERICARDIAL EFFUSION

IVC not collapsing with respiration

Right-heart catheterization

- PA pressure = 90/38 (mean 60 mmHg)
- Mean RA pressure = 16
- PCWP = 9
- CI 3.5 = L/min/m2 by TD (2.7 by Fick).
- Refused termination again – (some risk to mother also)
- Initiated on sildenafil; cannot use ERA
- Need to anticipate further worsening during pregnancy
- IV epoprostenol (Veletri) initiated
- Admitted to ICU for remainder of pregnancy
- Sildenafil increased to 60 mg q8h
- Cautious diuresis
Two weeks later (25 weeks):
▪ RV improved on Veletri, pericardial effusion smaller
▪ Epigastric discomfort – not eating – TPN initiated
▪ Platelets 100K to 60K  ?EPO / ?SLE / ?HIT / ?HELLP
▪ Complement values low (C3 45 mg/dL; C4 < 10 mg)
▪ Steroids added to hydroxychloroquine
▪ Total placenta previa; C-section planned at 30 – 32 weeks
▪ Mild crackles - ?atelectasis
At 27 weeks:
- Veletri increasing, I/O negative, RV further improved
- Patient's anxiety / stress increasing
- Increasing bilateral crackles (Veletri dose at 24 ng/kg/ml)
- O2 requirement increased from 2 to 6L O2 by nc

Dilemma: Improved RV function, but worsening crackles / hypoxemia
- Edema, +/- atelectasis?
- Infection? (no suspicious source, no fever, stable WBC)
- PVOD? PCH?
- Veletri weaned from 24 to 20 ng/kg/min
- O2 requirement stable off at 6L
- Delivery now planned for 28 weeks

Massive Pulmonary Edema and Death After Prostacyclin Infusion in a Patient With Pulmonary Veno-occlusive Disease

Patient was admitted to the hospital for treatment of pulmonary hypertension associated with PVOD. She had been receiving prostacyclin therapy for several months. On admission, she developed severe pulmonary edema and died shortly thereafter. The pathogenesis of this complication remains unclear, but it highlights the potential risks associated with the use of prostacyclin in patients with PVOD.

4 days before delivery.....
Delivery at 28 weeks

- Delivery moved up to 28 weeks
- ECMO catheters in place / intubated by cardiac anesthesia
- INO initiated
- Platelets down to 43K - transfused
- Continuous PA pressure monitoring
- RV watched very closely (mild dilation / hypokinesis)
- C-section completed – babies ok

- Bleeding → 2U PRBC
- Uterine atony → hysterectomy, continued Hgb drop
- Back to OR, explored, oozing no definite site
- To IR, anterior distribution of internal iliac arteries embolized
- To ICU on vent on 100% FIO2

Pulmonary edema worse...
Chest CT done....
After delivery

- Temp to 101 post-op
- Transaminases increased to 500 / 1000, creat 0.5 to 1.6
- Blood from ET tube x several days
- Prob DIC, platelets dropping – cryo / platelets transfused
- Veletri weaned to 14 ng/kg/min – RV stable
- No proof of infection – empiric antibiotics
- Multifactorial ST – intravasc vol depeletion, anxiety, drugs

- Diuresis, Veletri, low dose iNO, milrinone
- Aggressive pulm toilet, careful increase in PEEP
- O2, PEEP, weaned / extubated 2 weeks after delivery
- LFTs, creatinine normalized
- Plan to continue sildenafil, wean off Veletri and transition to selexipag

The New England Journal of Medicine

Original Article

Selexipag for the Treatment of Pulmonary Arterial Hypertension

Olivier Sitbon, M.D., Richard Channick, M.D., Kelly M. Chin, M.D.,
Aline Frey, Pharm.D., Sean Gaine, M.D., Nazzares Galié, M.D.,
Hossein-Andeschi Ghofieni, M.D., Marius M. Hoeger, M.D., Irene M. Lang, M.D.,
Ralph Press, M.D., Lewis J. Rubin, M.D., Lila Di Scala, Ph.D., Victor Tapson, M.D.,
Igor Adzerikho, M.D., Jinming Liu, M.D., Olga Moiseeva, M.D., Xiaofeng Zeng, M.D.,
Gerald Simonneau, M.D., and Valerio V. McLaughlin, M.D.,
for the GRIPHON Investigators®

December 24, 2015

Abstract
Challenges with this case

- Twin pregnancy
- Improved RV function on Veletri...
- But worsening hypoxemia / pulm edema ?PVOD / PCH
- How to handle Veletri / sildenafil
- Atelectasis, ? pneumonia
- Progressive thrombocytopenia, creatinine LFT↑.
- Role of SLE in low platelets and non-PAH lung issues
- Anorexia, abdominal pain requiring TPN
- Bleeding at delivery
- Transfusions / hypotension / fluid balance issues
- Hypoxemia post delivery but RV function stable
- Use of positive pressure / PEEP with abnormal RV
- Stress/ anxiety

CASE 2: 72 year-old woman - dyspnea x 1 year

- PMH = childhood asthma – resolved
- March 2015: F/u PCP visit. Improved, but still dyspneic with stairs / inclines. Told to lose 20 lbs
- May 2015: Lost 8 lbs, still dyspneic. Repeat CXR minimal LLL scar / infiltrate / scar, given albuterol (no wheezing).
- July 2015: Dyspnea clearly worse. Referred to pulmonary.
- Sept 2015: Pulmonary visit. Spirometry, CXR unremarkable. Referred to cardiology, and nutrition / dietary clinic.

Echo revealed PH 10 months after onset of dyspnea

- Oct 2015: Seen by cardiology. Echo revealed “PH.”
- Nov 2015: Amlodipine initiated for “PH.”
- Jan 2016: No improvement. Bilateral ankle edema. Started on sildenafil 20 mg q8h.
- Family researched PH online. Made appt with PH center.
- Feb 2016: PH center evaluation. With detailed questioning, she noted that regarding pneumonia admission a year before, she had no fever, no cough, only DOE, left chest pain and abnormal CXR.
- ANA 1:40, BNP 266 pg / mL, 6MW 381m
CHEST RADIOGRAPH
“Pneumonia” admission 1 year prior

Poor inspiration, infiltrate / effusion…

CTEPH - Mosaic perfusion…
Estimated PA pressure 81 mm Hg
RV mildly enlarged, mild hypokinesis

Right heart cath performed...

Estimated PA pressure 78/36 (mPAP = 50 mm Hg)
Right atrial pressure (RAP) = 12 mm Hg
PCWP = 8 mm Hg
Cardiac index = 2.4 L/min/m²
Vasodilator challenge not done

- CTA – mosaic perfusion, webs, post-stenotic dilation
- Abnormalities very distal, patient evaluated by experienced PH center, deemed inoperable
- Riociguat initiated, titrated up
CONCLUSIONS:

- Have low threshold for early referral to a PH center
- The diagnosis (cause and severity) must be confirmed with right-heart catheterization
- A VQ scan should always be done to exclude the possibility of CTEPH
- Aggressive therapy should always be considered in PAH in pregnancy
- Parenteral prostanoid therapy may be beneficial, even in suspected or proven PVOD / PCH, but caution is advised.

Thank you...
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