Medications of Pulmonary Hypertension

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Disclosures

- I have no disclosures. Any mention of brand names is for academic purposes and/or related to market availability and is not meant to endorse any product.

Brief review of pathophysiology of PH

- Pulmonary hypertension vs. Pulmonary arterial hypertension
Brief review of pathophysiology of PH

• Diagnostic criteria
  • RHC
  • MPAP > 25
  • PAWP < 15

• WHO Classes
  • Class I – PAH
  • Class II – PH with LH dysfunction
  • Class III – PH associated with lung disease
  • Class IV – PH d/t thrombotic/embolic disease
  • Class V – Miscellaneous

(McLaughlin et al., 2009)
(Revised WHO classification of PH)

• Basic science
• Symptoms
  • Dyspnea
  • LE swelling
  • Chest discomfort
  • Fatigue
• Complications
Calcium channel blockers

- nifedipine, diltiazem, amlodipine
- Recommended only if responsive to vasodilators
- Decreased MPAP, PVR, can help R heart
- Lack of evidence for extended-release formulations
- If doesn't respond symptomatically, need to move on to other therapies
- Used in PH for vasodilator effect

Phosphodiesterase 5 Inhibitors

- Sildenafil, Tadalafil
- PO
- cGMP causes vasodilation, but is degraded to GMP by phosphodiesterase type 5
- PDE 5 inhibitors block this degradation
- These two have different dosing!
- Used in PH for vasodilator effect
**Endothelin receptor antagonists**

- Bosentan, macitentan, ambrisentan
- PO
- Endothelin-1 causes vasoconstriction. In PAH, too much!
- Hepatotoxicity, teratogenicity, edema, h/a, nasal congestion

(Ataya, Cope & Alnuaimat, 2016)

**Prostanoids**

- Prostacyclin synthetics
- Epoprostenol, treprostenil, iloprost, selexipag
- IV, PO, SQ, Inhaled
- Jaw pain: a common side effect, GI symptoms, h/a, flushing

(Ataya, Cope & Alnuaimat, 2016)

**Soluble guanylate cyclase stimulator**

- Riociguat
- Oral TID dosing
- Stimulation of guanylate cyclase causes vasodilation
- H/a, dyspepsia

(Ataya, Cope & Alnuaimat, 2016)
Some others

- Anticoagulation
- Diuretics
- O2
- Digoxin

(Bishop, Meara, & Khouri, 2012)

Common side effects

- HYPOTENSION, syncope
- Headache
- Jaw pain
- Dizziness
- GI complaints
- Teratogenic

(Archer, Cope, & Alnuaimat, 2016)
(Archer, Wai, and Williams, 2015)

Monitoring parameters

- Orthostatic vital signs
- Side effects – are they tolerable?
- Systemic BP
- 6-minute walk test
- NYHA Functional Classification I-IV
Problems to consider

- Support system
- Ability to manage complex medication regimen
- Education level
- Access to tertiary care center
- Indwelling catheters

You see this person in your ER... now what?

- Take the time to find out what they are on and for how long. Who provides their PH care?
- Don’t stop an infusion! Don’t flush the line!
- Don’t change the dosing weight
- Do call the patient’s PH center, or consult your own expert if you have one.
- Examine central line for complications

Expert care team

- RN
- MD
- NP
- Prior authorizations – extremely time consuming
Palliative care

- Remember: symptom management
- Anyone with a chronic illness and sequelae should have some access/discussion of goals, symptoms
- Maybe a referral, maybe patient can be managed by pulmonologist

Lung transplantation

- For PAH or for underlying lung disease
- Transplant is exchanging one set of problems for another
- Almost always needs to be bilateral if for PAH – why do you think this is?
- Induction and transplant surgery are complicated
- For patients on IV prostacyclin, careful planning for OR transition is necessary
References


