Clarify transition in goals of care to comfort-oriented approach

☐ Clarify involvement of PAH specialist (usually Pulmonologist or Cardiologist) given their long standing relationship.
☐ Address spiritual and psychosocial concerns of patient and/or family.
☐ Consider benefits/burdens of the prostanoid infusion as symptom management in end-of-life care.
☐ Discuss the possibility of continuing versus withdrawing prostanoid infusions and address potential concerns:
  - increased symptom burden with withdrawal of prostanoid infusion
  - fear of uncontrollable symptoms
  - perception of prostanoid infusion as “life-line” for many years
  - benefits/burdens of wean versus abrupt discontinuation
☐ Reassure that focus of care will be aggressive symptom management.

Develop end-of-life care plan including discontinuation of prostanoid infusion

<table>
<thead>
<tr>
<th>Stable patient</th>
<th>Critically-ill/ Decompensated patient</th>
</tr>
</thead>
<tbody>
<tr>
<td>• Able to be at home OR stable enough to leave the hospital.</td>
<td>• Unable to leave the hospital setting (likely in an ICU)</td>
</tr>
<tr>
<td>• If decision has been made to wean as opposed to abruptly discontinuing the prostanoid infusion, consider the following:  - fast wean: decrease 20% per day  - slow wean: decrease by 2 ng/kg/min every other day to weekly  - if patient has dyspnea not controlled by other means, consider increasing to previously tolerated dose for 24 hrs, then re-initiate wean</td>
<td>• On aggressive life-supportive measures such as:  - mechanical ventilation  - hemodynamic support</td>
</tr>
<tr>
<td>• Re-evaluate with patient regarding whether to continue wean or discontinue abruptly</td>
<td>• Prevent initiation of other medically futile interventions</td>
</tr>
<tr>
<td>• Continue oral agents as tolerated**</td>
<td>• Abruptly discontinue the prostanoid infusion  - no need to wean  - consider pre-treatment with opioids to prevent respiratory distress with discontinuation of infusion</td>
</tr>
<tr>
<td>- may discontinue abruptly when no longer tolerated</td>
<td>• Discontinue other aggressive life-supportive measures based on best practice principles to minimize symptom burden and individualized for the patient’s clinical condition</td>
</tr>
</tbody>
</table>

TREAT SYMPTOMS AGGRESSIVELY
Use non-pharmacologic and pharmacologic interventions

Considerations for transition to hospice:

- All medications for PAH are expensive and will likely NOT be covered by Hospice
- Use home supply of PAH medications when possible
- Ask pharmaceutical companies for “compassionate care” use of PAH medications to cover cost
- May need inpatient hospice care for aggressive symptom management with discontinuation of prostanoid infusions

Medications:

- Prostanoid infusions: Epoprostenol (Flolan®), Treprostinil (Remodulin®)
- Oral agents: Endothelin-receptor antagonists (e.g. Bosentan, Tracleer®), PDE-5 Inhibitors (e.g. Sildenafil, Revatio®)
References:


