

Treatment Strategies for Pulmonary Arterial Hypertension in the Young

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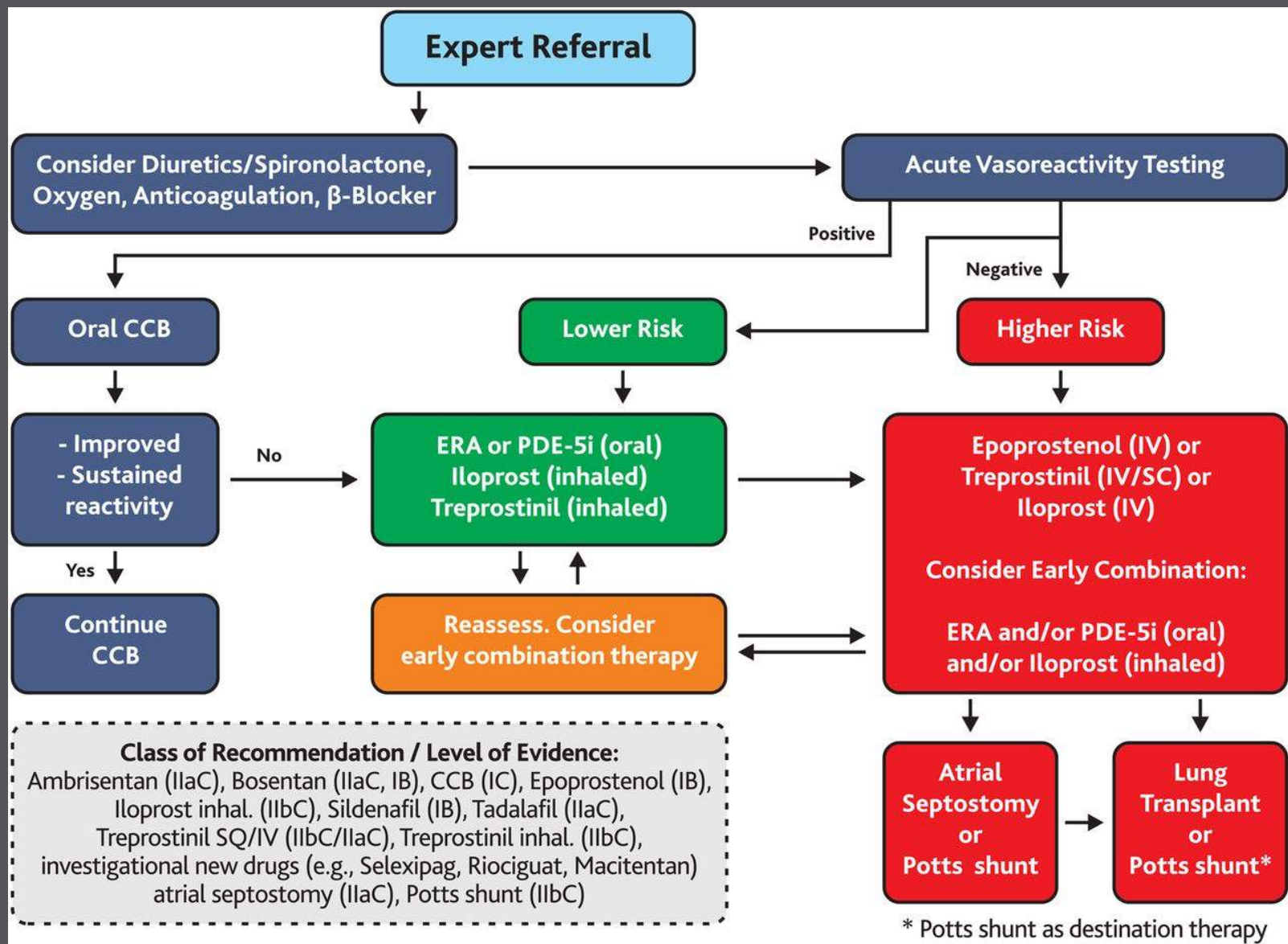


Disclosure

- I have no financial interests to disclose
- I will be discussing off-label uses of medications
 - I AM NOT CONDONING OR DISCUSSING
RECREATIONAL USES OF THESE MEDICATIONS

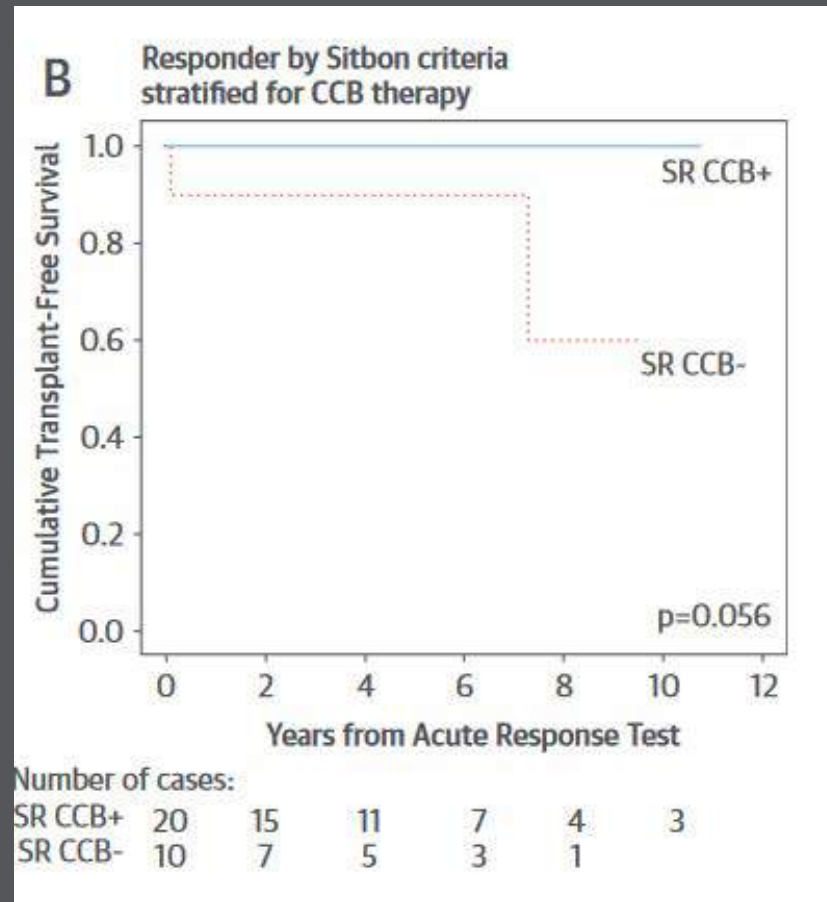
Outline

- Framework for treatment
- Medications
- Interventional options



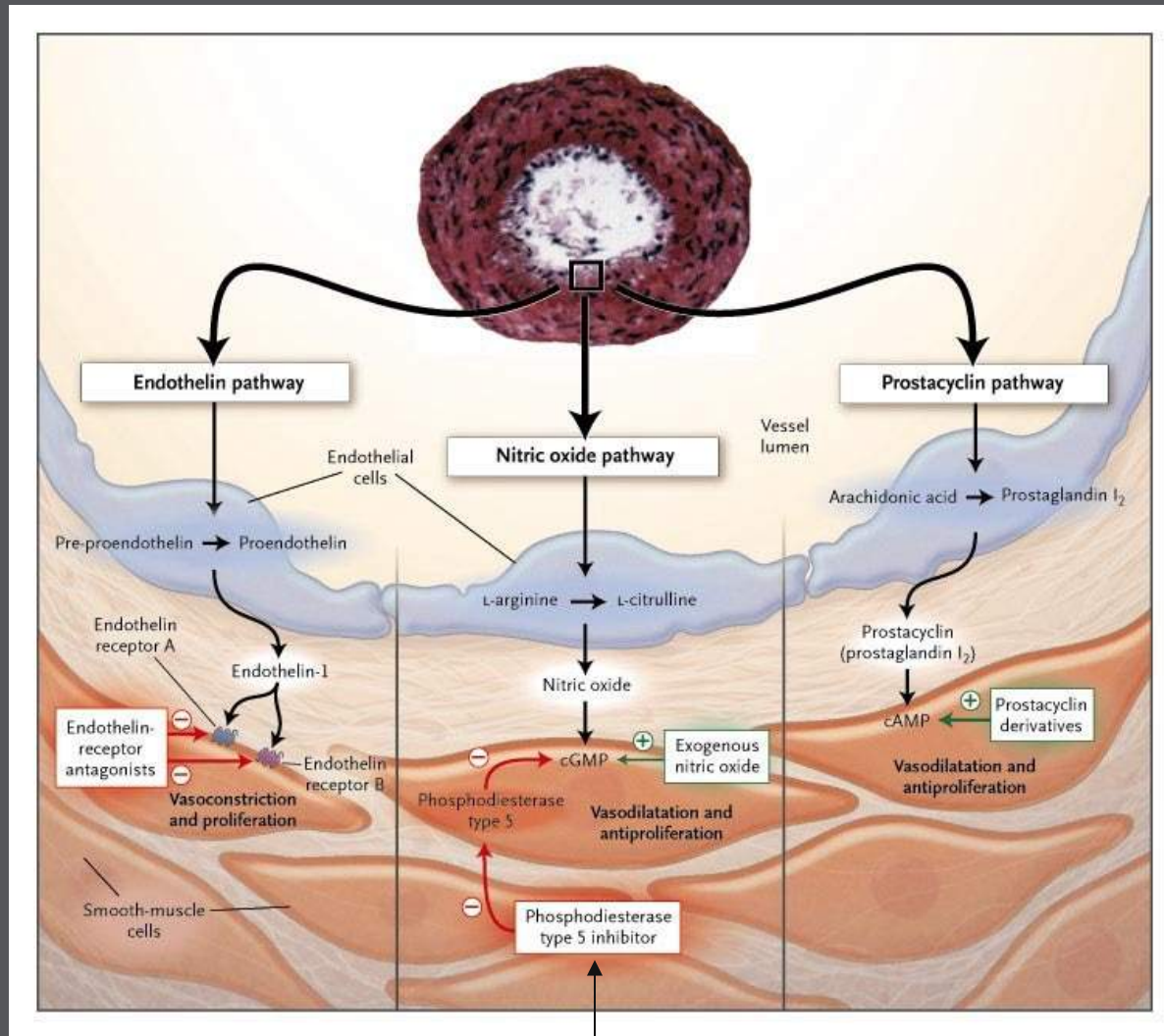
Hansmann, BMJ Hear
2016

Importance of CCB Therapy



Douwes *et al.* JACC.
2016

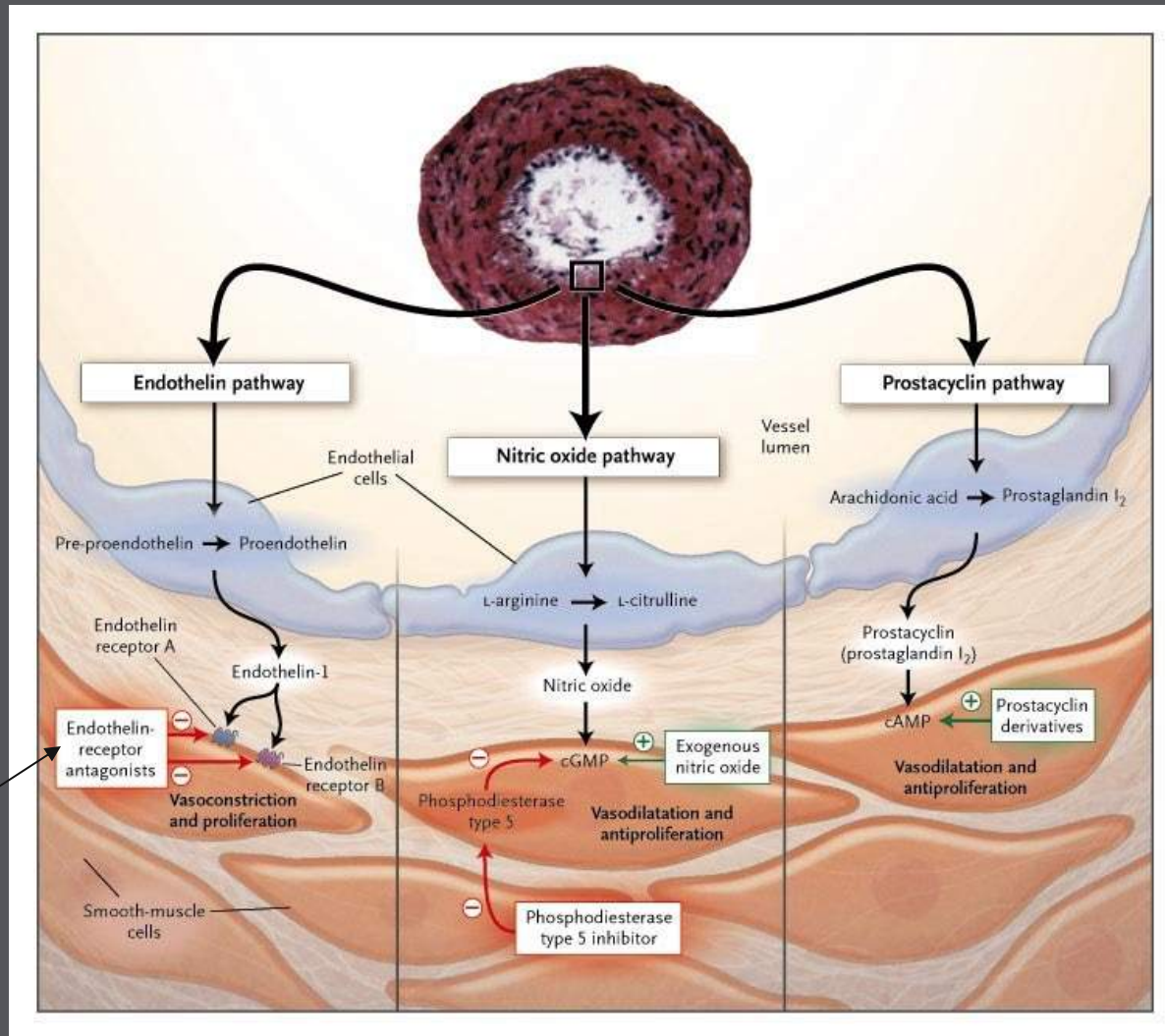
PAH Pulmonary Vasodilators: NO pathway



iNO, Sildenafil, Tadalafil, Riociguat

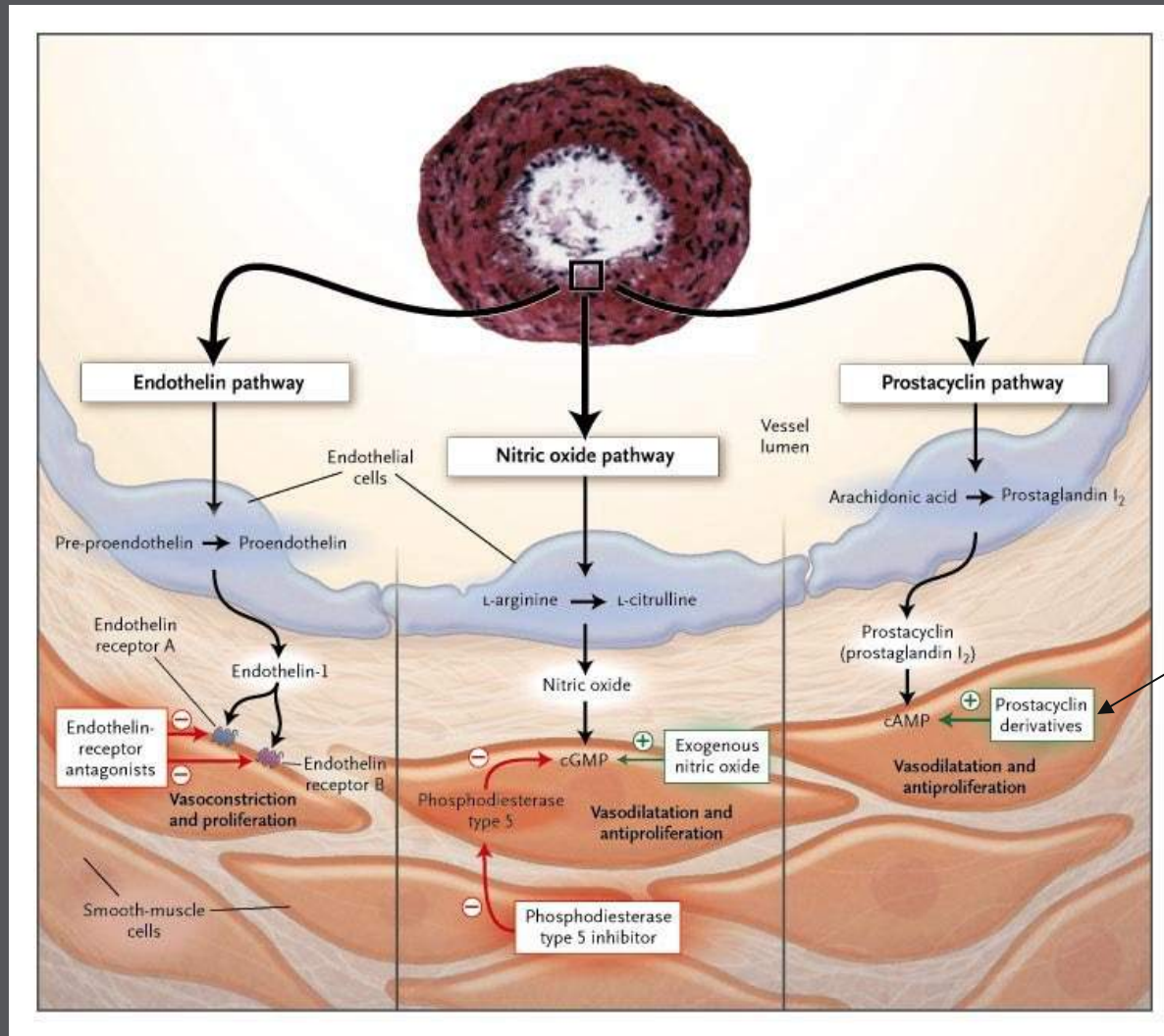
Humbert *et al.* NEJM
2004

PAH Pulmonary Vasodilators: ERA



Humbert *et al.* NEJM
2004

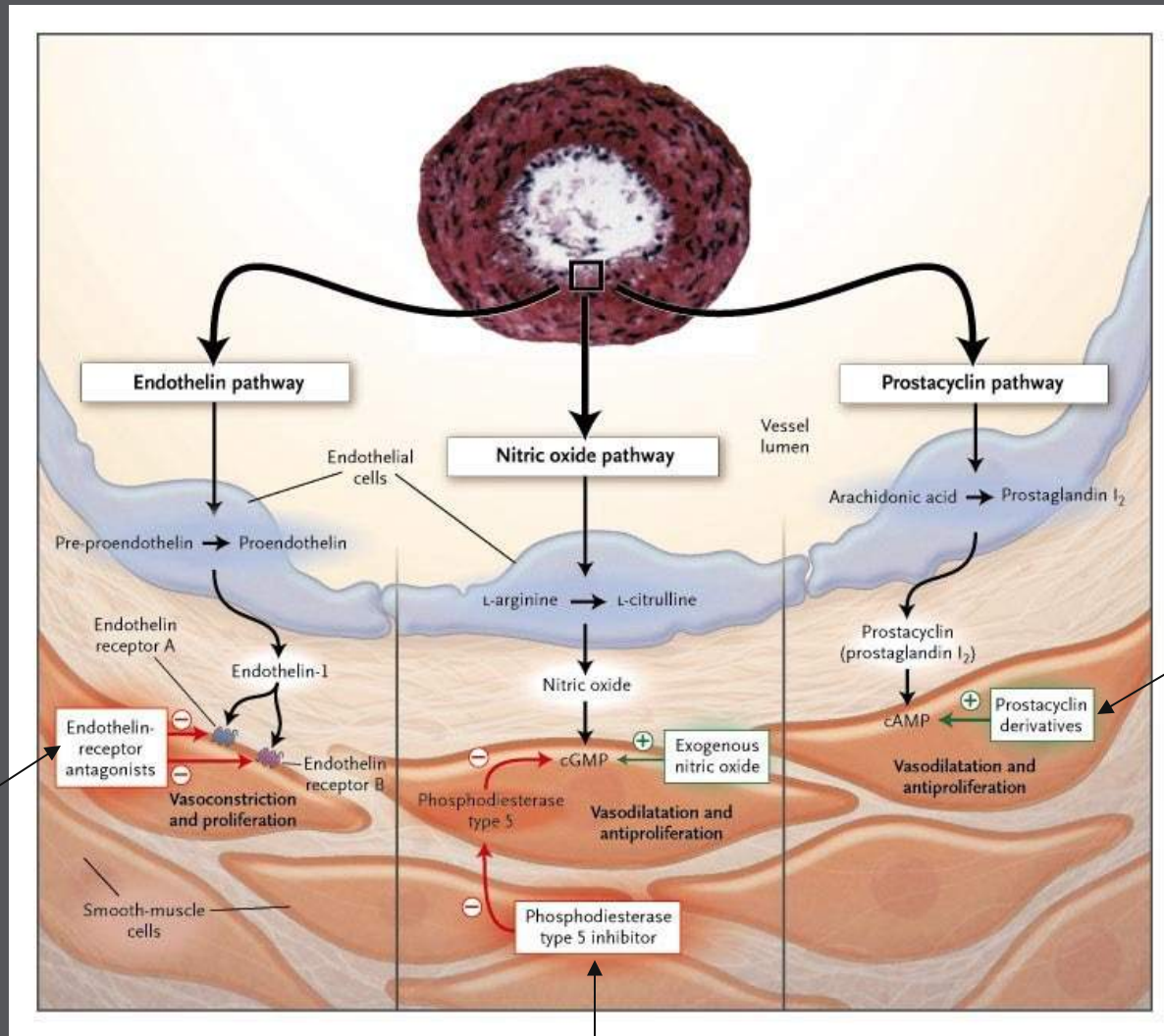
PAH Pulmonary Vasodilators: Prostacyclins



Epoprostenol
Treprostinil
Selexipag

Humbert *et al.* NEJM
2004

PAH Pulmonary Vasodilators:



Epoprostenol
Treprostinil
Selexipag

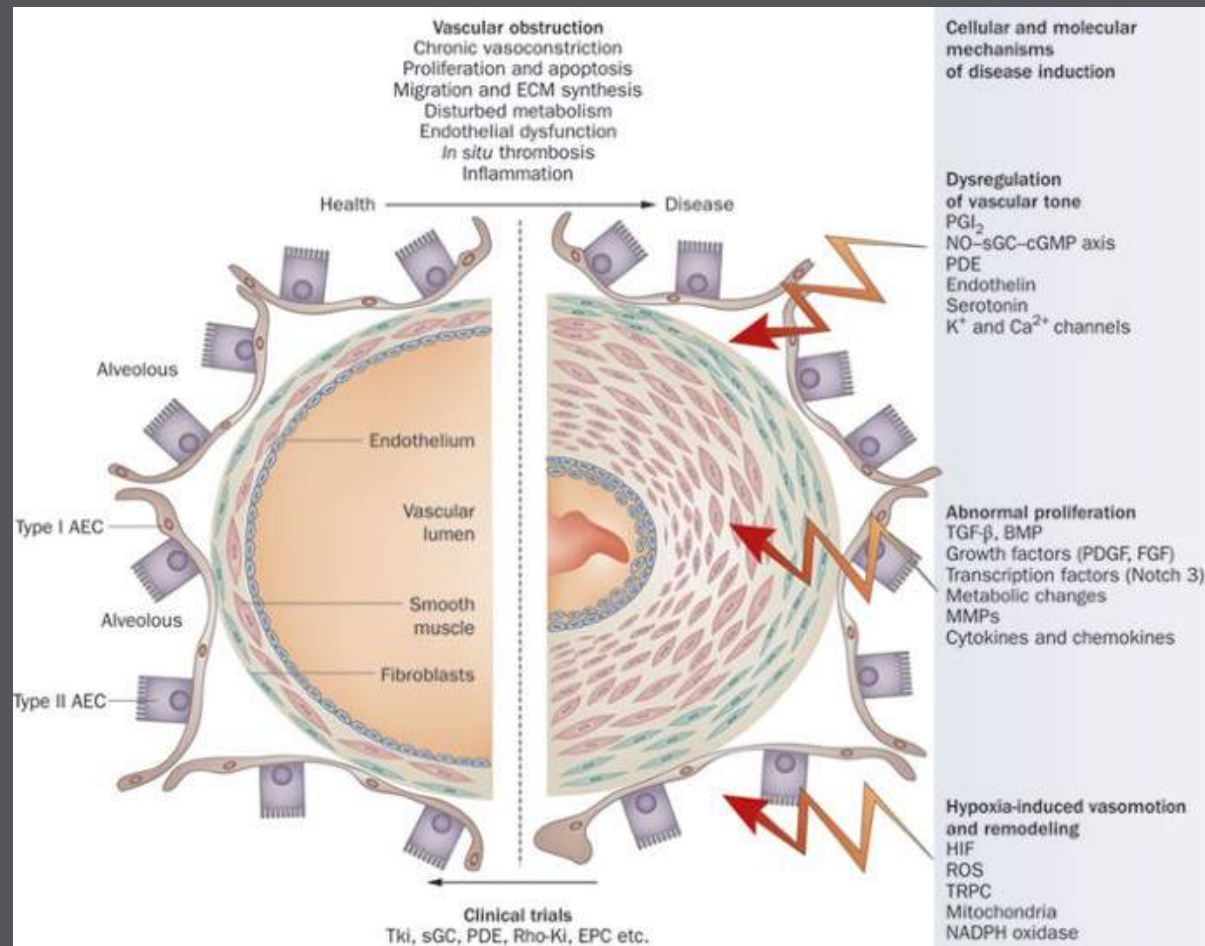
iNO, Sildenafil, Tadalafil, Riociguat

Humbert *et al.* NEJM
2004

Sotatercept

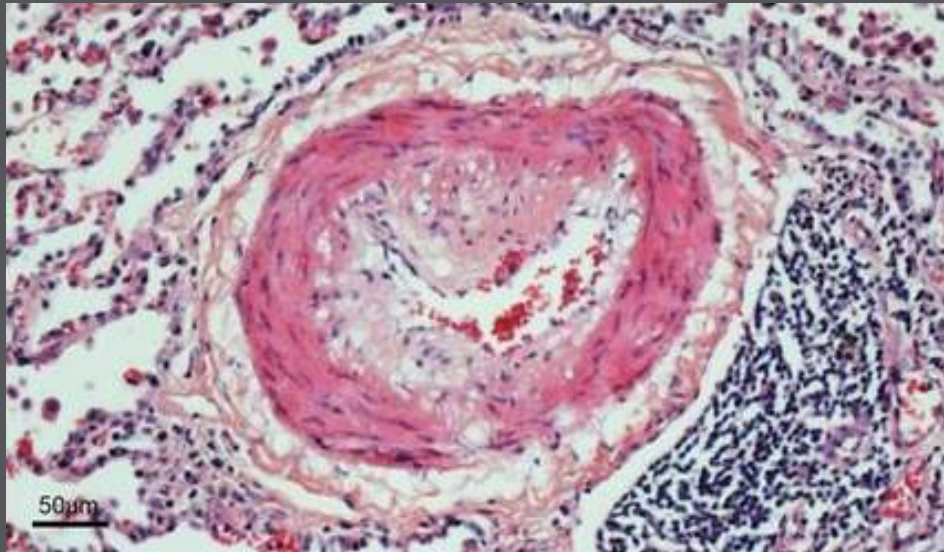
- Restoring balance of BMPR2 pathway
- Adult study: decrease PVRi and NT-proBNP
 - Mild symptom improvement
- Pediatric study: early stages

Molecular Pathways of PH

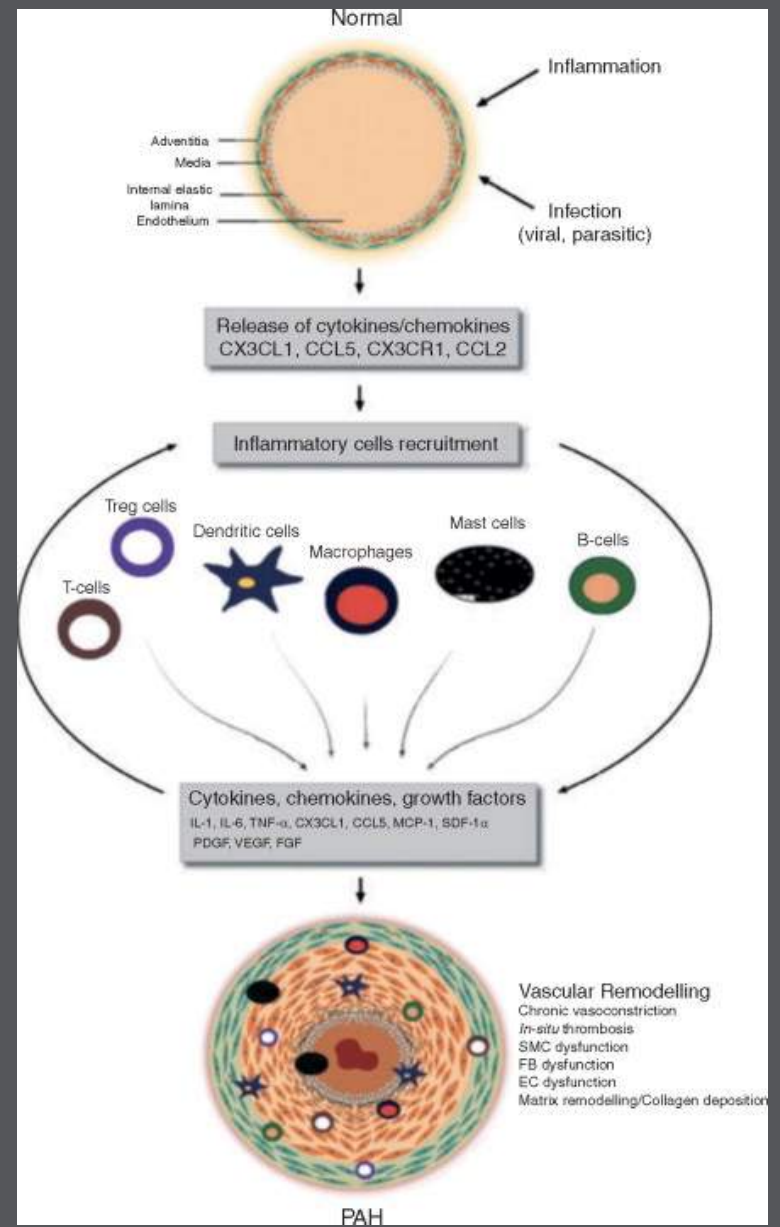


Schermuly *et al.* Nature Reviews 2011.

Inflammation in PAH



Rabinovitch, *Circulation Research* 2014



Pullamsetti, *Clinical Microbio and Inf Dis.* 2011

Steroids to Treat Pulmonary Hypertension

Dexamethasone reverses monocrotaline-induced pulmonary arterial hypertension in rats

L.C. Price^{*,#,§,+}, D. Montani^{*,#,§}, C. Te N. Gambaryan^{*,#,§}, M-C. Chaumais^{*,#}, L.S. Howard[§], I.M. Adcock[†], S.J. Wort

Prednisolone Ameliorates Idiopathic Pulmonary Arterial Hypertension

To the Editor:

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Glucocorticoids for treating paediatric pulmonary hypertension: a novel use for a common medication

Manish Aggarwal, Ronald Mark Grady

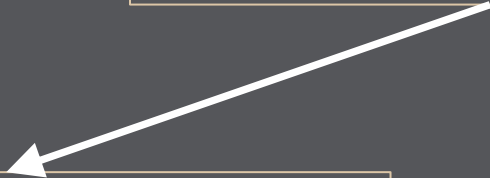
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Worsening

Pulmonary vascular disease



↑ Pulmonary vascular
resistance



RV Dilation/Hypertrophy

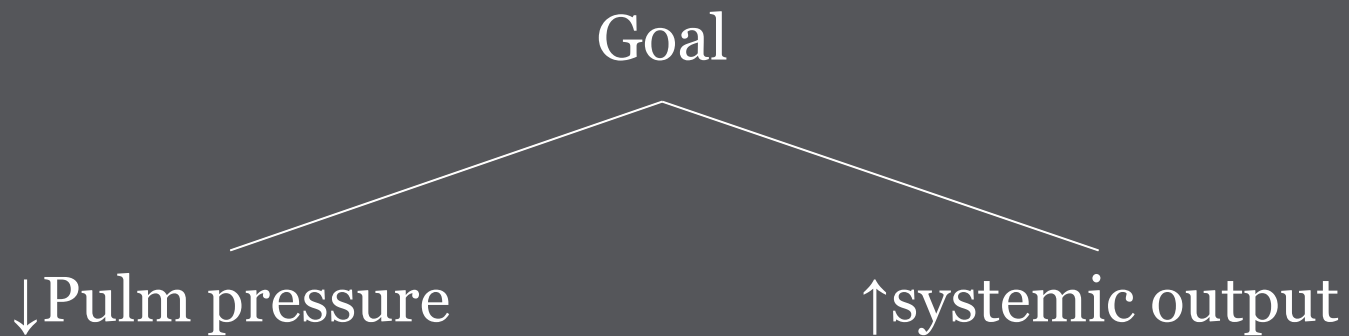
RV Failure



Death

RV Failure

- ↑Pulmonary pressure impairs RV function
- ↓RV function → ↓pulmonary and systemic output

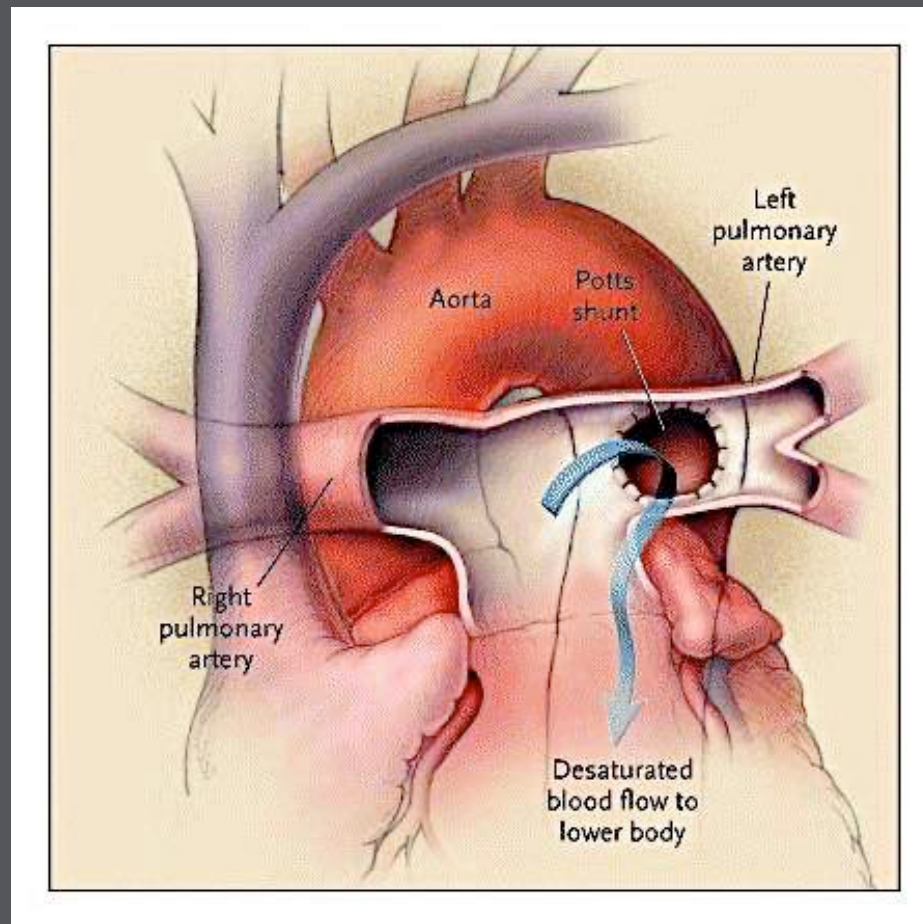


Atrial Septostomy = Diastolic Pop-off



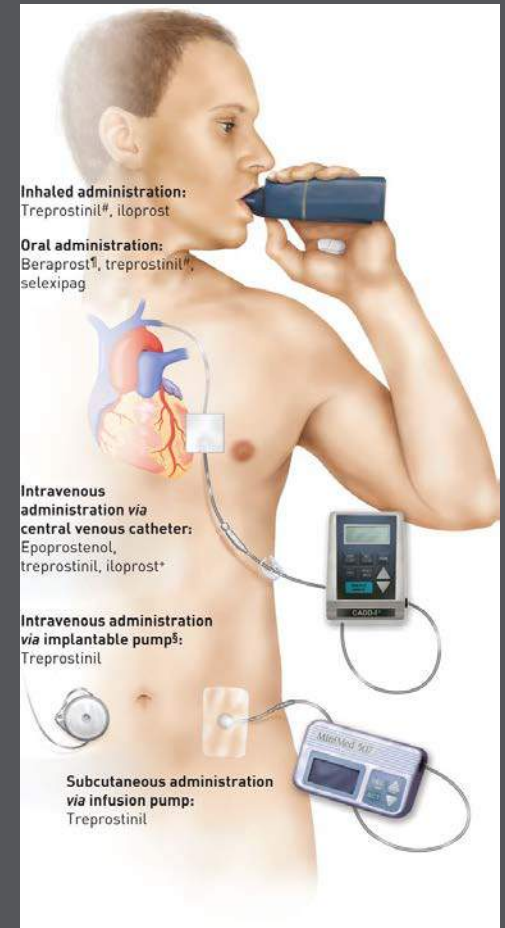
Occlutech AFR

Systemic Potts Shunt

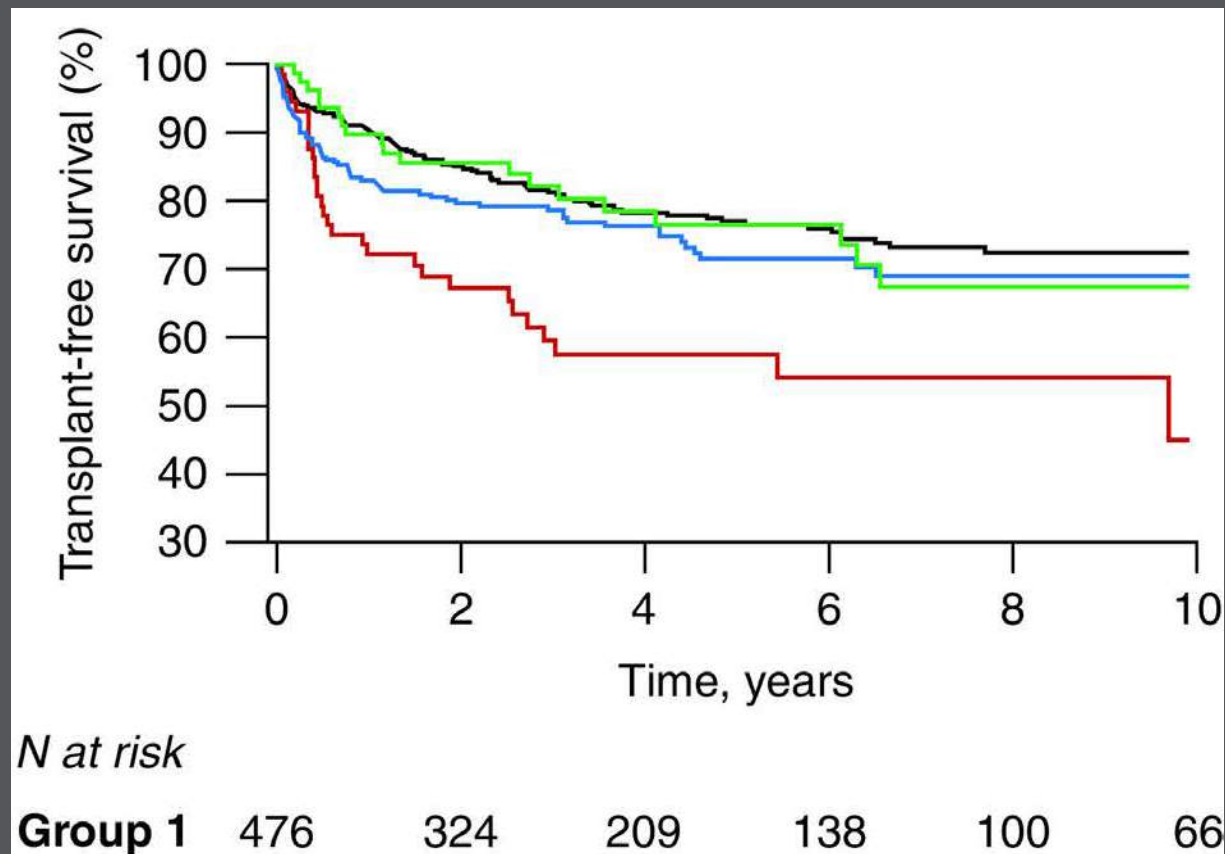


Blanc *et al.* NEJM 2004.

Costs



Treatments but not Cures

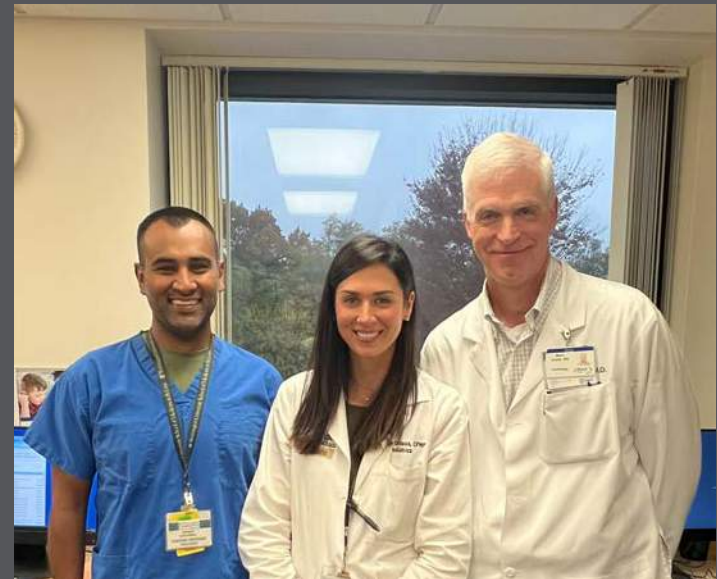


Constantine, *et al.*
AJRCCM 2022

Conclusions

- Medication options: CCB, NO pathway, ERA, prostacyclins
- Similar medications for ~20 years
- Alternative medications are on horizon?
- Interventional techniques including atrial septostomy and Potts shunt are available

Thank you!



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