Pathophysiology
PAH is defined as a mean pulmonary pressure > 30 mmHg during exercise or equivalent, or PAP/artP > 50%. New classification [1] in Venice 2003 defined:
Three metabolic pathways could be modulated in clinics: the nitric oxide, the endothelin and the prostacyclin.

New drugs
Nitric oxide and derived drugs
Inhaled nitric oxide (NO) is a selective pulmonary vasodilator easily used in OR. 10 to 20 ppm is the most usual dose. Corvasal®, sildenafil (Viagra®, inhibitor of phosphodiesterase V) are the most popular and effective agents.

Bosentan
Bosentan (Tracleer®) is a new anti-endothelin inhibitor. It has a few hemodynamic effects, but a long term anti proliferative action. Primary pulmonary hypertension or related to chronic embolism, HIV, vascularitis are good indications.

Prostacyclin
Prostacyclin (PGI2, Flolan® or beraprost) is a selective pulmonary vasodilator with endothelial and antiplatelet short term action. It is the best choice in non responder and severe patients, or young patients with primary pulmonary hypertension.

Inotropes
Milrinone is a short term phosphodiesterase III inhibitor, more frequently used than enoximone. Indication is right ventricular failure with preserved arterial systemic pressure or combined with norepinephrine.
Levosimendan, a calcium sensitizer, without initial loading dose (to avoid hypotensive effect) is effective at 0.05 to 0.1 µg/kg/min and has prolonged beneficial effect, after 5 days.

Anesthesia
Ketamine, morphine and propofol don’t change hypoxic vasoconstriction, NO2 or pentothal decrease. All the data are not available for inhaled anesthetics, but sub MAC-desflurane and sevoflurane seem to be safe.