Southern African Journal of Anaesthesia and Analgesia 2018; 24(3)(Supplement 1)

South Afr J Anaesth Analg
ISSN 2220-1181 EISSN 2220-1173

FCA 2 REFRESHER COURSE

An approach to the pregnant patient with pulmonary hypertension

E Mostert

Department of Anaesthesiology, Chris Hani Baragwanath Academic Hospital, University of the Witwatersrand
Correspondence to: estie254@gmail.com

Introduction

Pulmonary hypertension (PH) in pregnancy is rare, with an incidence of 0.7 per 100 000 pregnancies.[1] In PH there is a constant increase in pulmonary arterial pressure and pulmonary vascular resistance (PVR). There is a decreased prostacyclin and nitric oxide (NO) production and increased thromboxane production.[2] This leads to failure of the right ventricle and eventually death.[3] PH in pregnancy is associated with a high mortality rate, previously found to be 30–50%, but decreased in recent reviews (16–25%).[4-6] The mortality rate for women with Eisenmenger's syndrome is 30–70%.[2]

Most literature still advocates for women of childbearing age with PH not to fall pregnant, or terminate a pregnancy early in the first trimester.[4,6-8] However, some patients will only be diagnosed during pregnancy or still choose to become pregnant. These patients require a multidisciplinary team approach to their management regarding pregnancy and delivery.

Definition and classification

PH is defined as a mean pulmonary arterial pressure (mPAP) 25 mmHg or more at rest as diagnosed by right heart catheterisation.[9] PH is regarded as severe if the mPAP is > 45 mmHg.[10] Table I is a summarised clinical classification of PH. Patients with associated left heart disease (Group 2) are characterised by pulmonary wedge pressures > 15 mmHg, or post-capillary PH, while all the other groups have pressures < 15 mmHg, or pre-capillary PH. The term pulmonary arterial hypertension is used for patients in Group 1.[6] The histopathological changes that occur differ between the groups but can include medial hypertrophy, proliferation of the intima, fibrosis, thrombotic lesions and venous thickening.[8] The concern with PH is the development of right heart failure, and depending on the onset of disease, it can be acute or chronic.[2]

Table I. Summarised clinical classification[8]

1. Pulmonary arterial hypertension
   - 1.1 Idiopathic
   - 1.2 Heritable
   - 1.3 Drug and toxin-induced
   - 1.4 Associated with
     - 1.4.1 Connective tissue diseases
     - 1.4.2 HIV
     - 1.4.3 Portal hypertension
     - 1.4.4 Congenital heart disease
     - 1.4.5 Schistosomiasis
     - 1.4.6 Chronic haemolytic anaemia
   - 1.5 Persistent pulmonary hypertension of the newborn
   - 1.6 Pulmonary veno-occlusive disease/pulmonary capillary haemangiomas
2. Pulmonary hypertension due to left heart disease (systolic or diastolic, valvular disease)
3. Pulmonary hypertension due to lung disease and/or hypoxia (COPD, interstitial lung disease etc)
4. Chronic thrombo-embolic pulmonary hypertension
5. Pulmonary hypertension with unclear and/or multifactorial mechanisms (haematological, systemic, metabolic)

Pregnancy and labour physiology

The physiological changes during pregnancy are not tolerated well by patients with PH. The most significant changes occur in the cardiovascular and respiratory systems. The blood volume increases by 40–50%. This peaks at 20–32 weeks gestation.[11] Heart rate and stroke volume also increase and lead to an increased cardiac output. A further increase in cardiac output happens during labour and delivery (25 –50%). In the immediate postpartum period the cardiac output can be more than 80% of prelabour values due to autotransfusion from uterine contraction.[2,6] The complex haemodynamic changes that occur during labour and delivery are usually not tolerated well in PH, causing acute strain on the right heart and pulmonary vasculature.[4]

In healthy women, the pulmonary circulation undergoes vasodilatation, to accommodate the increase in blood volume, but in PH the pulmonary circulation is unable to cope with the haemodynamic changes due to pulmonary vascular remodelling. This leads to an increase in pulmonary pressures as the cardiac output increases. The right ventricle may be unable to increase the cardiac output and the patient may present with dyspnoea, heart failure and syncope.[6] In the presence of Eisenmenger's syndrome, the fixed pulmonary vascular resistance and lower systemic vascular resistance (SVR) cause a worsening in right-to-left shunt and hypoxia.[6]

Thrombotic events may be aggravated during pregnancy (hypercoagulable state) and can worsen pulmonary and peripheral vascular thrombosis.[6]

Risk of death in these patients is highest during delivery and in the postpartum period.[12] Pulmonary vascular resistance increases and right ventricular contractility may decrease in the immediate postpartum period. Should the preload also decrease, patients with pulmonary arterial hypertension may have cardiovascular collapse. Other mechanisms that can lead to sudden death include: arrhythmias, pulmonary embolism or stroke from intracardiac shunts.[12]

Timing and mode of delivery

The timing and mode of delivery for optimum outcomes are still controversial. Women with severe PH have a significantly higher risk for preterm delivery. A planned early delivery (usually before 34 weeks), while clinically stable, contributes to better maternal outcomes. Caesarean section (CS) delivery has become the predominant mode of delivery, although vaginal delivery is not necessarily contraindicated in mild PH.[4]

Vaginal delivery is usually associated with less bleeding, decreased risk of infection and lower thromboembolic risk. The sudden changes in haemodynamic parameters associated with CS are also avoided. However, a long duration of labour can be detrimental to the mother in terms of an increase in cardiac output and venous return associated with pain.[13]

The advantages of a planned CS include: daytime scheduling, avoiding the risk of urgent CS during hours where less experienced healthcare providers are available. Reports suggest better outcomes with planned CS and the use of regional anaesthesia.[4]

Labour analgesia

Early effective analgesia is essential to maintain SVR and PVR balance to avoid sympathetic stimulation from pain. If placement of a labour epidural is contraindicated due to anticoagulation, a low-dose remifentanil infusion or patient-controlled analgesia are suitable alternatives.[2]

Preoperative evaluation

Symptoms of PH include fatigue, dyspnoea, angina and syncope. Syncope is usually associated with a poor prognosis.[14] Signs of PH may be subtle and can include: a prominent jugular a-wave, palpable left parasternal lift, loud pulmonary component of the second heart sound, mid-systolic ejection murmur, diastolic murmur (pulmonary regurgitation) and a pan-systolic murmur (tricuspid regurgitation – associated with a prominent jugular venous v-wave, hepato-jugular reflux and a pulsatile liver). Features of right ventricular failure include ascites, peripheral oedema, distension of the jugular veins, pulsatile hepatomegaly and a right ventricular third heart sound.[15]

The following tests are useful as a mortality predictor:

1. Functional capacity (NYHA or WHO)
2. Exercise capacity (unencouraged six-minute walk test)
3. Haemodynamics (severity of right ventricular dysfunction)
4. Echocardiographic parameters[15]

Echocardiography can aid evaluation of estimating pulmonary artery pressures, left and right ventricular function and size, abnormalities of valves and presence of intracardiac shunts.[14] Although right heart catheterisation is described in the literature as necessary to confirm the diagnosis of PH,[9,12,14] this is not necessarily done in a resource-constrained environment. For planning the management of PH, it is important to diagnose and optimise the underlying cause of PH, including obstructive sleep apnoea, COPD, recurrent thromboembolism, cardiomyopathy and valvular heart disease.[14]

Treatment of PH will depend on the underlying cause, patient's haemodynamic status and disease severity. Anticoagulation therapy is usually prescribed for women with pulmonary arterial hypertension and in chronic thromboembolic disease. The use of anticoagulation therapy in women with portal hypertension or Eisenmenger's syndrome is controversial because of an increased bleeding risk.[4,6]

When there are clinical signs of cardiac failure, diuretics (furosemide and hydrochlorothiazide), digoxin and fluid restriction are recommended.[2,6]

Treatments specifically for PH include: calcium channel blockers, endothelin receptor antagonists (ERA), phosphodiesterase-5 inhibitors and prostacyclin analogs. ERAs are teratogenic, and should not be used in pregnancy.[4] Treatment for PH should be continued up to and including the day of surgery. Warfarin should be stopped timeously and patients bridged with low-molecular weight heparin. [2]

Further investigations include: full blood count, metabolic panel, coagulation studies and a preoperative arterial blood gas. An ECG to detect signs of right-sided strain or ischaemia should be done.[2] Cardiovascular review with special investigations (echocardiography as a basic investigation), right heart catheterisation and CT angiogram may be necessary.

Intraoperative management

The best method of anaesthetising women with PH for CS is controversial and data is limited and mostly retrospective from case reports.[16,17] For patients with moderate to severe PH, spinal anaesthesia is contraindicated due to the sudden changes in SVR and preload.[2]

Anaesthetic and haemodynamic goals

1. Maintain preload
2. Avoid increase in PVR: prevent hypoxia, hypercarbia, acidosis and pain
3. Maintain SVR (Decreased SVR reduces cardiac output due to a "fixed" PVR)
4. Avoid myocardial depressants, maintain myocardial contractility
5. Maintain sinus rhythm[2]

Monitoring

1. Arterial line – indicated for most patients
2. Central venous access – placement should be done cautiously to avoid inducing arrthythmias
3. Cardiac output monitoring – TEE or pulse pressure variation monitoring should be considered in severe PH cases.[2] Case reports suggest the use of pulmonary artery catheters.[16,18]

Regional anaesthesia

This offers an effective method of pain control for labour and delivery (vaginal and CS). Combined spinal-epidural has been described in the literature, but the use of a graded epidural is more common. The use of single-shot spinal anaesthesia is contraindicated as this leads to decreased venous return and preload, leading to systemic hypotension. A graded epidural provides more control over the level of sympathetic blockade, speed of onset and haemodynamic stability, however the risk of hypotension is still present.[4]

General anaesthesia

The use of general anaesthesia is still controversial in patients with PH, and its use has been linked to an increased mortality as compared to regional anaesthesia.[5] However, it may be indicated for emergency CS, when regional is contraindicated or with an insufficient block and in patients with active heart failure.

There is limited literature identified to support a specific technique for general anaesthesia. As with regional anaesthesia, general anaesthesia should be administered with caution to avoid vasodilation and protect the haemodynamic status of the patient.[2,4]

For induction, standard agents have been used successfully, despite a decreased SVR, (propofol and succinylcholine for rapid sequence induction). Etomidate can also be considered as it is associated with less haemodynamic effects. Blunting of the intubation response with high-dose short-acting opioids and lignocaine is important; however, the use of opioids can cause respiratory depression in the neonate. The use of volatile inhalational agents should be with caution, as it can also decrease SVR and have negative inotropic effects.[4]

Oxytocin should be used with caution, as it can lead to systemic hypotension and increase PVR.[2]

Assuming euvolaemia, hypotension should be managed with inotropes.

Dobutamine or agents like levosimendan or milrinone can be used as intravenous inotropic therapy. This can cause systemic vasodilation and hypotension that can be managed with phenylephrine or noradrenaline. Phenylephrine and noradrenaline can increase PVR.[2]

Inhaled NO is a pulmonary vasodilator that can be used, however, it requires special equipment for inline administration in the circuit, and it is not available everywhere.[2] Extracorporeal membrane oxygenation (ECMO) can be used where available as rescue therapy for PH crisis. Inhaled prostacyclin can also be used.[4]

Ventilation should be aiming to avoid hypoxia, hypercarbia and lung hyperinflation. Low tidal volume ventilation using low PEEP levels and increased respiratory rate to avoid hypercarbia are advocated.[2]

Postpartum management

It is important to monitor PH patients in a critical care environment during the postpartum period for a few days postpartum. Fluid status should be monitored and fluids used with caution. Treatment for PH should continue and may need to include inhaled NO or prostacyclin. Phosphodiesterase-5 inhibitors should be restarted immediately after delivery. Anticoagulation therapy is recommended for most postpartum patients, especially for patients with chronic thromboembolic disease.[17] The cardiovascular changes start returning to prepregnancy levels soon after delivery, however, this can take up to six months to complete.[4]

Conclusion

Patients with PH have a significant morbidity and mortality risk. The management of these pregnancies should be with a multidisciplinary team approach. Literature is limited to case reports, with no randomised control trials to evaluate which anaesthetic technique is the most effective. Anaesthetic management should be tailored to each patient, according to disease severity and planned mode of delivery. Both graded epidural and general anaesthesia have been used with success. Most deaths occur in the postpartum period, so careful monitoring should be done in an intensive care setting.

References

1. Knight M, McClymont C, Fitzpatrick K, et al. On behalf of UKOSS. United Kingdom Obstetric Surveillance System (UKOSS) Annual Report. National Perinatal Epidemiology Unit 2012.
2. Weitzel N. Perioperative management solutions for pulmonary hypertension. An update and review. Available: www.ucdenver.edu/academics/colleges/medicalschool. Accessed 25/04/2018.
3. Sahni S, Palkar AV, Rochelson BL, et al. Pregnancy and pulmonary arterial hypertension: A clinical conundrum. Pregnancy hypertension: An International Journal of Women's Cardiovascular Health. 2015;5(2):157-164.
4. Viktorsdottir O. Pulmonary hypertension in pregnancy and anesthetic implications. Current Anesthesiology Reports. 2015;5(1):82-90.
5. Bédard E, Dimopoulos K, Gatzoulis MA. Has there been any progress made on pregnancy outcomes among women with pulmonary arterial hypertension? European Heart Journal. 2009;30(3):256-265.
6. Pieper PG, Lameijer H, Hoendermis ES. Pregnancy and pulmonary hypertension. Best Practice and Research Clinical Obstetrics and Gynaecology. 2014;28(4):579-591.
7. Hemnes AR, Kiely DG, Cockrill BA, et al. Statement on pregnancy in pulmonary hypertension from the Pulmonary Vascular Research Institute. Pulmonary Circulation. 2015;5(3):435-465.
8. Pieper PG, Hoendermis ES. Pregnancy in women with pulmonary hypertension. Netherlands Heart Journal. 2011;19(12):504-508.
9. Pritts CD, Pearl RG. Anesthesia for patients with pulmonary hypertension. Current Opinion in Anesthesiology. 2010;23(3):411-416.
10. Yahya M, Saleem MY, Chandra M, et al. Anaesthesia for a patient with severe pulmonary hypertension coming for Cesarean section: A case report. Sch J Med Case Rep. 2015;3(1):7-9.
11. Bassily-Marcus AM, Yuan C, Oropello J, et al. Pulmonary hypertension in pregnancy: critical care management. Pulmonary Medicine. 2012;2012:9.
12. Smith JS, Mueller J, Daniels CJ. Pulmonary arterial hypertension in the setting of pregnancy: a case series and standard treatment approach. Lung. 2012;190(2):155-160.
13. Olsson KM, Channick R. Pregnancy in pulmonary arterial hypertension. European Respiratory Review. 2016;25(142):431-437.
14. Ortega R, Connor CW. Intraoperative management of patients with pulmonary hypertension. advances in PH. 2013;12(1):18-23.
15. Madden BP. Pulmonary hypertension and pregnancy. International Journal of Obstetric Anesthesia. 2009;18(2):156-164.
16. AlBackr HB, Aldakhil LO, Ahamd A. Primary pulmonary hypertension during pregnancy: A case report. Journal of the Saudi Heart Association. 2013;25(3):219-223.
17. Bonnin M, Mercier FJ, Sitbon O, et al. Severe pulmonary hypertension during pregnancy mode of delivery and anesthetic management of 15 consecutive cases. Anesthesiology. 2005;102(6):1133-1137.
18. Lin DM, Lu JK. Anesthetic management in pregnant patients with severe idiopathic pulmonary arterial hypertension. International Journal of Obstetric Anesthesia. 2014;23(3):289-290.