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Obstetric anaesthesia in rheumatic heart disease – a unique perspective from the Top End

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INTRODUCTION

Rheumatic heart disease (RHD) is a complex manifestation of social inequity. It is a life-long burden and, in Australia, affects predominantly young First Nations Australians. It is endemic in the Northern Territory (NT), with a rate of disease 26 times higher than the rest of Australia.[1] Despite intensive primary and secondary prevention, it is not a disease of the past, with episodes of acute rheumatic fever (ARF) continuing to increase in the NT. Of those suffering an episode of ARF, 50% will develop rheumatic heart disease within 10 years, of which one-third will be severe. With episodes of ARF occurring almost exclusively in children, this translates to the majority of subsequent heart disease occurring in those aged 15-44. Females are consistently over-represented in this population leading to a disproportionate burden of disease in women of childbearing age.[2]

As the tertiary referral centre in the Northern Territory, the Royal Darwin Hospital is in the unique situation of managing a relatively high volume of obstetric patients with rheumatic heart disease, often from the most remote locations in Australia. Up to 2-3% of First Nations women who become pregnant in the NT have some form of RHD, and many have had previous valve interventions.[3] This presents not only the challenge of managing valvular heart disease in pregnancy and the peripartum period, but also in a way that addresses different concepts of health and the additional complexities of distance, language, and culture.

CARDIOVASCULAR CHANGES IN PREGNANCY AND LABOUR

Pregnancy is a dynamic physiological process with significant alterations to the cardiovascular system. These changes are driven by the endocrine effects of progesterone, the addition of the parallel placental circulation, and the response to flow-metabolism coupling in the presence of increased metabolic demands. There is up to a 50% increase in cardiac output (CO), primarily due to an increase in blood volume, heart rate (HR), and a steady decrease in systemic vascular resistance (SVR).[4] A physiologic anaemia of pregnancy occurs due to an increase in blood volume in excess of the concurrent increase in red-cell mass. This reduces blood viscosity and resistance to blood flow.[5] The predominant haemodynamic state is hypervolaemic, hyperdynamic, and vasodilated. The net effect is decreased systolic blood pressure (SBP). These changes peak in the 28th week of pregnancy.

At the time of first and second-stage labour (unassisted by anaesthesia), there is a sudden additional increase in HR and CO due to the sympathetic activation from pain and autotransfusion of up to 500 ml of blood from each uterine contraction. Valsalva manoeuvres during pushing decrease the preload and increase the afterload.[6] Further autotransfusion occurs in the third stage from the placental circulation and unloading of aortocaval compression. CO then reduces precipitously to pre-pregnancy levels within 24-48 hours.

These changes are often poorly tolerated in patients with rheumatic heart disease, particularly those with stenotic valvular lesions and pulmonary hypertension, who struggle to cope with sudden increases/decreases in the circulatory volume, diastolic filling, SVR, and CO. Anaesthetic interference, such as epidural analgesia, will temper the more acute changes, while other interventions, such as rapid fluid administration or the induction of a general anaesthetic, will add an extra level of complexity.[7]

MATERNAL AND FOETAL OUTCOMES IN RHD

Most of the evidence on outcomes in pregnancy in RHD comes from overseas, from low-income countries, where RHD accounts for a greater proportion of obstetric cardiac disease. One of the largest prospective datasets is from the registry of pregnancy and cardiac disease (ROPAC), which published outcomes for a series of 390 patients with rheumatic mitral disease in pregnancy in 2018. In this series, the mortality rate was 1.9%; however, nearly 50% of patients with severe mitral stenosis (MS) and 23% with severe mitral regurgitation (MR) developed heart failure. In addition, severe MS was an independent risk factor for adverse foetal outcomes.[8] Foetal outcomes in maternal cardiac disease are poor due to reduced uteroplacental flow from obstructive left heart disease. This translates into low birthweight, preterm delivery, and higher rates of neonatal intensive care admissions and stillbirth.[9] A recent 2020 systematic review of 12 studies had similar findings, with mitral stenosis being associated with a 3% mortality rate, 38% rate of heart failure, and independently associated with adverse foetal outcomes.[10] This is consistent with several other recent publications where severe MS and pulmonary hypertension were independent risk factors for maternal morbidity.[9,11,12] It should be noted that these outcomes are more pronounced in low income, low health literacy settings where antenatal care is compromised, and resources are limited.[13,14]

Australian data, limited in numbers, indicates a lower overall mortality rate but still significant morbidity and adverse effects on the foetus. For example, recent retrospective studies in Western Australia (WA) and the NT recorded no maternal mortalities, while two prospective studies recorded a mortality rate of 0.3-0.4%.[9,13,15,16]

A retrospective review of RHD in pregnancy in the NT from 2010 to 2019 indicated at least 13 cases of RHD in pregnancy being managed per year, with up to 25% of patients in the modified World Health Organization (mWHO) class III and IV risk categories (Figure 1).[17] There were no maternal or neonatal mortalities in the study. Ninety-one per cent of patients identified were from remote areas, and interestingly there was a relatively high rate of postpartum haemorrhage (24%) relative to national data (5-15%).[15] In our experience, the total number of cases and representation of critical events are likely to be significantly underestimated due to limitations in retrospective data collection. To our knowledge, we can recall specific patients in the time-period of this review who suffered considerable morbidity, and this review should not give false confidence in the management of these complex patients.

RISK STRATIFICATION OF RHD IN PREGNANCY

Many risk-scoring systems are available to classify the burden of cardiac disease in pregnancy (CARPREG I, CARPREG II, ZAHARA).[12,18,19] Of those available, the mWHO classification[20] is considered to be better at predicting maternal outcomes.[21,22] It classifies maternal risk into four categories, with level I representing the lowest risk of a maternal cardiac event (2.5-5%) and level IV representing the highest (40-100%). For level IV patients, pregnancy is not recommended before surgical or interventional management of their cardiac condition.

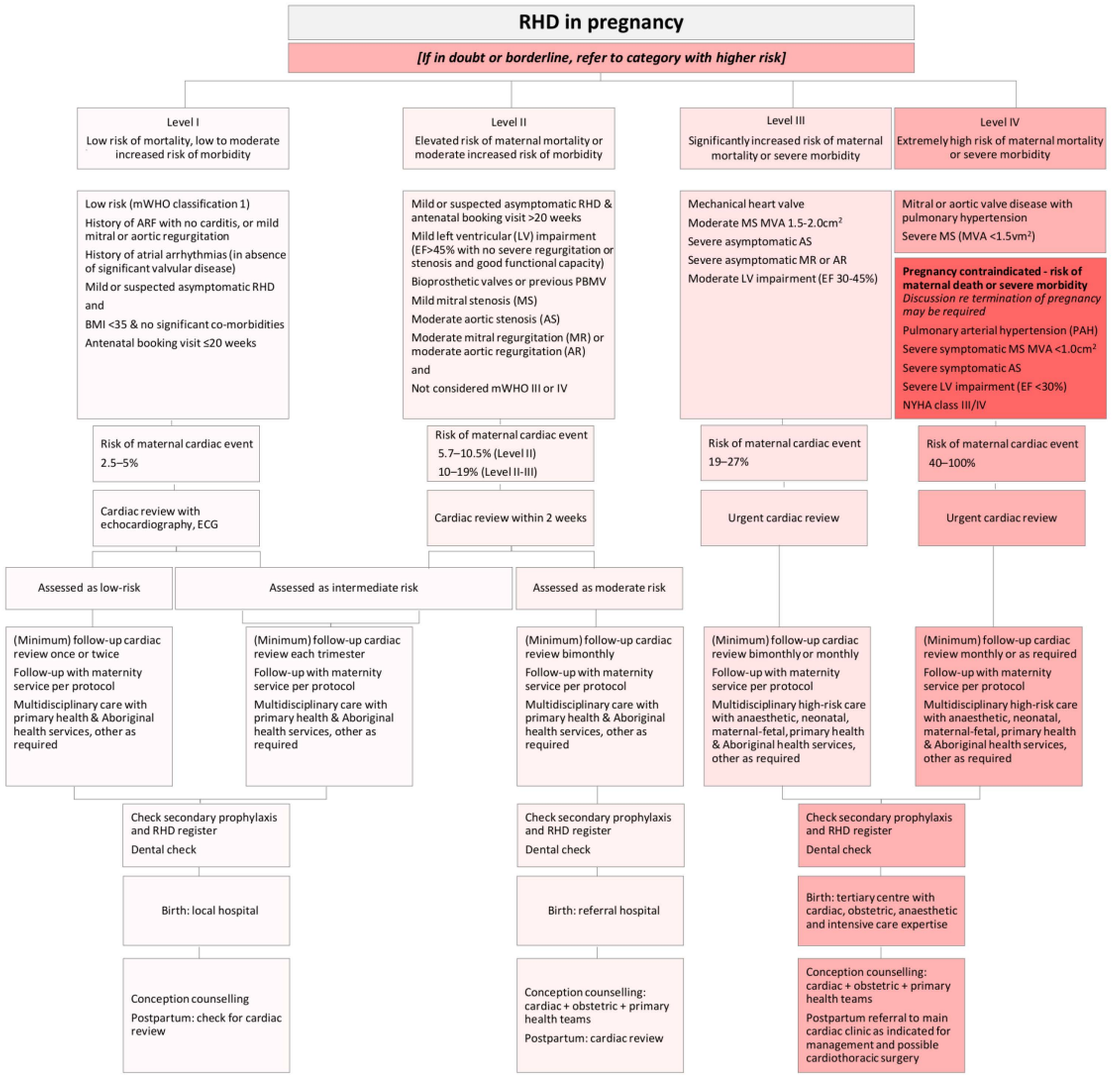
The mWHO classification applies to congenital disorders, aortopathies, and advanced procedures such as heart transplantation. RHD Australia (an Australian government co-ordination unit aimed at controlling RHD/ARF) has adapted the risk stratification to focus on the valvulopathies seen in rheumatic heart disease, particularly the risk posed by differing severities of mitral valve stenosis and pulmonary hypertension.[23]

Management during pregnancy and planning for delivery is guided by the mWHO classification (Figure 1). Level I patients can generally be managed in the community, with a single cardiology review and a local delivery.[21] Patients with a level II classification require follow-up with bimonthly cardiology reviews and delivery in a larger centre. Patients classified as level III and IV (where termination has been declined) need monthly cardiology review and accommodation within easy access to a tertiary centre, where delivery should occur. All patients require some level of multi-disciplinary team (MDT) planning for delivery, which should involve shared

decision-making with the patient and their family. For patients classified as level III and IV, the MDT involves an obstetrician and maternofetal medicine specialist, neonatologist, cardiologist, intensivist, and anaesthetist with assistance from interpreters and Indigenous liaison officers (42% of First Nations patients use English as their second language).[24] The expansion of online platforms and telehealth in the NT has dramatically improved the ability to facilitate these meetings and reviews, reducing unnecessary travel and keeping women in community for longer.

Figure 1. Classification of RHD in pregnancy

Reproduced from the 2020 Australian guideline for prevention, diagnosis and management of acute rheumatic fever and rheumatic heart disease[25]



Regarding risk stratification, two important clinical caveats are observed.

Firstly, the increase in cardiac output secondary to hypervolaemia will increase the transmitral valve gradient, often without associated changes in mitral valve area (by planimetry).[26] Therefore, it is not unusual for patients with a mitral valve area (MVA) >1.0cm^2 to have a transmitral mean gradient >10mmHg later in pregnancy. For this reason, the mWHO classification considers MVA <1.5cm^2 in pregnancy as severe, rather than using a transmitral gradient classification system. If a mean pressure gradient (MPG) classification system is used, a patient with a MPG >10mmHg in the first trimester of pregnancy is likely to be at a higher risk of complications compared to a patient who starts with a lower gradient and develops an MPG>10mmHg later in pregnancy.[27]

Secondly, the unique weather environment of the Top End of the NT is one of the greatest physiologic stress tests available to anaesthetists and perioperative physicians. Some Top End patients will arrive at the hospital following a pregnancy spent entirely in ≥30°C heat, 80% or more relative humidity, and with a daily step count to parallel professional athletes. Taking time to perform an appropriate functional assessment and understand the day-to-day life of patients often reveals an incredible physiologic reserve, which may weigh in on decision-making. For mWHO class III and IV patients, or where this physiological reserve is unclear, stress echocardiography or a six-minute walk test (6MWT) is frequently performed in the early third trimester to help stratify those who are more likely to decompensate. This is particularly useful to help overcome the limitations of NYHA classification in pregnancy, where shortness of breath can be multifactorial.

BEYOND CLINICAL PRIORITIES

While clinical priorities guide recommendations, for many patients the impact of non-clinical factors (cultural, social, financial) cannot be ignored. These may exacerbate any clinical risk if not appropriately addressed.[28] There are anaesthetists in the Top End currently engaged with communities to explore these factors, the differences in perception of healthcare, and the impact of Western medicine. While eagerly awaiting formal publication of this important work, we can only share our personal experiences and stories to give insight into these factors.

One of the biggest issues faced in the NT is the patient's disposition of where to deliver. For many First Nations patients, the severity of their condition means delivering away from their land and Country. High-risk patients will be transferred to Darwin in the third trimester. Without a local cardiac surgical service, patients will occasionally require transfer >3000km interstate if the need for cardiac intervention or mechanical circulatory support is anticipated. The transfer to Darwin, or especially interstate, can have significant social impacts. The transfer of a patient away from family, culture and Country can be likened to an international relocation, often while critically unwell and preparing to embark on the challenges of motherhood. Also, women may be too unwell to look after their child post-delivery, disrupting the maternal-neonatal connection. A small proportion of patients may require cardiac surgery, affecting their initial ability to breastfeed due to the sternotomy. The impact may be profound, with some women having to leave their babies interstate and return home alone in the face of this adversity. As clinicians, we must be aware of these flow-on effects when making transfer decisions.

We must be aware of how our perception of "good medicine" aligns with the patient's perceptions. Understanding resource limitations and staff expertise, the aim is to make every effort to support the pregnant woman's wishes. The importance of birthing on Country is paramount to many First Nations women, and when this is not possible, the concept should be applied as a metaphor. This requires designing services in collaboration with First Nations women to be inclusive, holistic, and culturally competent.[29,30]

PRECONCEPTION PLANNING

All women of childbearing age with significant rheumatic heart disease should have a pre-pregnancy assessment where patients can be counselled about the risk of pregnancy, have their cardiovascular status optimised, and further therapies planned, for example, mitral valvuloplasty. Where cardiac surgery is being planned, effective, long-term but reversible contraceptive therapies like an intrauterine device can be considered. Oestrogen-based contraceptives pose an added risk of thromboembolism and potentially increase the risk of mechanical valve thrombosis. Tubal ligation is discussed in cases where patients have completed their families.[25] Where pregnancy is contraindicated, patients are appropriately counselled, but the final decision will rest with the patient and should be respected. Cardiovascular disease and the inability to bear children can result in significant social stigma for the patient, and these topics need to be addressed in a multidisciplinary setting with social and cultural sensitivity.

ANTENATAL MANAGEMENT

Screening

Identification of RHD early in pregnancy allows for better risk management and time for intervention planning and implementation. Unfortunately, in low-resource settings, up to 25 per cent of RHD in pregnancy remains unidentified until unmasked by the haemodynamic changes of the second or third trimester.[14] Anecdotally, this is not infrequent in the NT, with patients occasionally presenting in heart failure as their first presentation. Recognising patients at risk starts in primary care, and the NT is currently running a pilot program called "The Two Heart Beats Study", whereby women from remote communities receive a screening echocardiogram with their first-trimester ultrasound. Access to echocardiography is challenging outside of a tertiary centre, and the emphasis is on providing primary care physicians with baseline skills to diagnose subclinical RHD.

Medical optimisation

RHD management in pregnancy involves balancing pharmacotherapy against relative risk(s) of teratogenicity. Hypertension, arrhythmias, anaemia and active infections, which may cause cardiac decompensation, can be addressed early. Heart failure and fluid overload can be managed with diuresis and rate control. This will help manage the risk of acute pulmonary oedema. Volume overload will result in an increase in transmitral gradients without a decrease in the valve area. Significant reductions in transmitral gradients can be achieved with well managed diuresis. A summary is included in Table 1.

Table 1. Medication management of RHD in pregnancy

THERAPIES	CHANGE IN PREGNANCY
Arrhythmia control	Beta blockers: continue (accept an increased rate of IUGR, neonatal hypoglycaemia). Digoxin is safe and routinely loaded.[31]
BP control	Aldosterone antagonists (AA), Angiotensin-converting enzyme inhibitors (ACEi) and Angiotensin II receptor blockers (ARBs) are discontinued due to teratogenicity. Alternative: hydralazine, labetalol, methyldopa as per non-cardiac pregnancy.[32]
Anticoagulation	Warfarin is teratogenic in the first trimester due to its effects on organogenesis, although both European and ASA guidelines allow warfarin up to 5 mg in the first trimester. RHD Australia also recommends changing to bi-daily LMWH only in the first trimester, then switching back to Warfarin from the 13th to the 35th week.[25,33,34] Change to heparin or Low Molecular Weight Heparin (LMWH) peripartum (around the 36th week) in preparation for neuraxial techniques. There is no consensus on optimal anticoagulation. Although LMWH has better foetal outcomes, it has higher rates of thrombogenicity. The fluctuating fluid states of pregnancy mean that weight-based dosing alone will not be accurate in achieving therapeutic levels so anti-Xa levels are recommended.

In remote communities, access to frequent testing can be problematic, and any change in therapeutic anticoagulation exposes the woman to an increased risk of thrombogenic complications. Local services aim to continue warfarin up until the 36th week, then switch to LMWH in the peripartum period. High risk patients (mechanical valves, thrombophilia or active thrombotic disease) are transferred to Darwin in the third trimester to have anticoagulation managed and monitored.

Surgical intervention

Percutaneous balloon mitral valvuloplasty (PBMV) is safe and results in good foetal outcomes in patients with severe stenotic lesions.[27,35,36] However, it carries the risk of acute severe mitral regurgitation and pericardial tamponade and is not always appropriate for patients with mixed valvular lesions. As there is no cardiothoracic surgical service in the NT to manage the complications of PBMV, patients needing cardiac intervention require interstate transfer to Adelaide.

DELIVERY

Planning

In the absence of obstetric indications for lower uterine segment Caesarean section (LUSCS), normal vaginal delivery is preferred due to lower rates of postpartum haemorrhage, infection, and thromboembolism.[37,38] Planned LUSCS can avoid emergent delivery and allow expert multidisciplinary input in the highest-risk cases. Spontaneous labour is reasonable for patients classified as mWHO II risk if they reside within close proximity to the hospital. For remote patients, this entails hotel-style accommodation, which can be provided in the weeks leading up to delivery. As mentioned previously, this often results in the displacement of many First Nations women from their support networks at a time of high stress. In patients classified as mWHO III and IV risk, the preferred approach is planned labour induction in the intensive care unit (ICU). This allows optimisation of timing, personnel, equipment, and resources with continuous monitoring during the dynamic stages of labour and the postpartum period. It also aids in the inpatient optimisation of volume status, rate control and management of anticoagulation before a neuraxial technique.

Anaesthetic considerations

Established RHD often presents as a mixed valvular disorder. Table 2 delineates the most common anaesthetic considerations for each valvular disorder. Haemodynamic aims are generally targeted to the most severe lesion; however, care must be taken not to underestimate the additive effects of moderate lesions in series (that is, aortic regurgitation and mitral regurgitation). Left-sided valve disease, stenotic lesions, and pulmonary hypertension carry the highest risk of patient morbidity and mortality.[20,27,39] Pulmonary hypertension due to obstructive left ventricular disease such as mitral or aortic stenosis is associated with a 30-56% risk of maternal mortality.[39] If these lesions are associated with right ventricular dysfunction or a poor functional score, the risk increases exponentially. In addition, the hypercoagulable state of pregnancy increases the probability of pulmonary emboli, which can, in turn, worsen pulmonary hypertension. A small pulmonary embolus may result in acute right ventricular failure in a patient who is maximally compensated.

Regurgitant lesions, in comparison, are generally well tolerated during labour, delivery and anaesthesia in the absence of heart failure and pulmonary hypertension.[12]

Many patients will have required previous valve interventions before becoming pregnant. In women of childbearing age, it is standard to offer a bioprosthetic valve replacement with subsequent surgery to convert to a mechanical valve after the completion of their family. Bioprosthetic valves do not require therapeutic anticoagulation like their mechanical counterparts but are at risk of structural degeneration and may need reoperation as early as within the first five years. This option is not always selected due to the requirement to return interstate for redo surgery. As a result, many women in the NT present in pregnancy with previous mechanical valve replacements, requiring therapeutic anticoagulation. It is not uncommon for patients to present in the NT with restenosis, obstruction of valve replacements, or failure of previous valve repairs.

Due to the complexity of valvular disorders and the rate of unexpected “moderate to severe” findings, all diagnostic transoesophageal echocardiography performed in Darwin is done in the operating theatre complex with a consultant anaesthetist.

Table 2: Anaesthetic goals and considerations in common RHD lesions and sequelae

Valvular lesion	Pathophysiologic processes	Anaesthetic goals (HR, preload, afterload, contractility)
Mitral stenosis	Fixed preload to the left ventricle (LV) impairs the ability to increase CO in demand to the effective vasodilated state of pregnancy. Hypervolaemia and tachycardia of pregnancy lead to an increased gradient across the mitral valve, left atrial (LA) dilation and increased pulmonary artery pressures. If severe pulmonary hypertension develops, right ventricular failure may also occur.	Heart rate: Prevent tachycardias and aggressively treat dysrhythmias (adequate analgesia, avoid terbutaline, defibrillation pads on). Normovolaemia: pre-emptive diuresis if evidence of pulmonary oedema or right ventricular failure before induction of labour. Preferential use of vasopressors over volume for hypotension management. Percutaneous balloon valvuloplasty (see surgical management below).
Aortic stenosis	Decreased SVR of pregnancy associated with the vascular changes and vasodilation of neuraxial anaesthesia decreases coronary perfusion pressures. Tachycardia decreases LV diastolic filling time and LV perfusion time. Often associated with LV diastolic function.	Gentle initiation of neuraxial analgesia and anaesthesia with concurrent management of the vasoplegia. Low threshold to treat haemorrhage with volume resuscitation. Transcatheter aortic valve replacement in specific cases may be warranted, safe, and effective.[40]
Regurgitant lesions – **MR and AR**	Forward regurgitant fraction is improved with a low SVR and a higher HR. Neuraxial will reduce the SVR which improves the regurgitant fraction. MR is associated with an enlarged LA and an increased risk of arrhythmias. APO may result from increases in LA and pulmonary pressures, particularly when there is an acute regurgitant change with increased SVR or acute ischaemia with chordae rupture.	Avoid increases in SVR. Avoid bradycardia – use ephedrine and chronotropic agents. Avoid arrhythmias – digoxin loading is commonly used in Darwin on patient admission. Acceptance of lower blood pressure may be an appropriate technique as patients will have increased forward flow. However, at some point hypotension will reduce placental circulation flow as this is a pressure-passive circulation.
Mechanical prosthetic valve	High risk of valvular thrombosis with hypercoagulable state of pregnancy.	Vitamin K antagonist continued in pregnancy and changed to LMWH in the peripartum period (as above). Higher risk of intra and postpartum haemorrhage. Consider early surgical management of postpartum haemorrhage. GA LUSCS if the anticoagulation cannot be stopped. Mechanical valve thrombosis is a life-threatening emergency (due to acute stenosis, regurgitation and heart failure) and requires urgent transfer to a cardiothoracic centre to provide (TOE-guided) thrombolysis or surgery to extract the clot.

Pulmonary hypertension/ right ventricular dysfunction	Increased circulating blood volume and venous return results in acute right heart dysfunction in the face of elevated pulmonary vascular resistance or poor RV reserve. Hypercoagulable state of pregnancy increases the risk of pulmonary emboli. Decreased SVR leading to decreased coronary perfusion to a hypertrophic RV reliant on diastolic perfusion. Placental blood autotransfusion postpartum may result in acute pulmonary hypertension and right ventricular failure.	Regular volume status tracking with diuresis where necessary. Maintain strict fluid balance and make haemodynamic decisions using invasive monitoring. Maintain right ventricular perfusion by maintaining SVR using vasopressor support. Invasive monitoring and pre-emptive vasopressor therapy is recommended when instituting neuraxial analgesia/anaesthesia. Careful incremental initiation of neuraxial analgesia and anaesthesia. Reduce PVR using nebulised milrinone or additionally inhaled nitric oxide or nebulised prostacyclin if intubated. Sildenafil is also a safe option in pregnancy. Prevent increases in PVR: avoid over sedation causing hypercarbia, administer supplemental oxygen. Limit oxytocic use. Oxytocin boluses in 0.5-1 unit increments with vasopressor support to offset hypotension. Ergometrine and carboprost are contraindicated as they can increase pulmonary pressures. Therefore, early surgical management is the mainstay of treatment of PPH. Under GA: Avoid over or under ventilation to maintain normocarbia and high intrathoracic pressure, and use TOE to monitor RV and cardiac output.

Monitoring

All parturients should have standard ANZCA monitoring, including a five-lead electrocardiogram to allow early detection of arrhythmias and subendocardial ischaemia.

Arterial line monitoring with or without pulse contour analysis is utilised to guide vasopressor management in the face of haemodynamic insults like neuraxial analgesia and postpartum haemorrhage. However, arterial waveform analysis has limited application in cases of rapid haemodynamic disturbances such as during a LUSCS.

The use of a central venous access line is indicated in high-risk cases where the use of vasoactive medications is expected; however, there is no proven utility of central venous pressure (CVP) monitoring in this patient population. Pulmonary artery catheters are generally not required.[41,42]

Defibrillation pads are placed pre-emptively in case of unstable arrhythmias, and fluid balance is monitored with an indwelling urinary catheter.

Peripartum sequential transthoracic echocardiography (TTE) can help guide management, and if the patient is intubated, a TOE provides more detailed information due to improved windows.

Continuous cardiotocography (CTG) for foetal monitoring is indicated prior to any intervention.

Anticoagulation

For patients on regular anticoagulation, Vitamin K antagonists have usually been ceased in the peripartum period and changed to LMWH. Therapeutic anticoagulation with LMWH must be ceased at least 24 hours before any attempted neuraxial technique.[43] A neuraxial technique may be required earlier than anticipated if spontaneous labour is induced by the insertion of the balloon for cervical ripening on the day prior to induction. This needs to be considered when deciding on the timing of cessation. Overall, therapeutic anticoagulation in the peripartum period will increase the risk of peripartum haemorrhage and needs to be weighed against the risk of valvular or systemic thrombosis, which is made worse by the hypercoagulable state of pregnancy.

Fluid management

Preload to the right ventricle is increased in the immediate postpartum period due to autotransfusion from the uterus and decompression of the IVC from the non-gravid uterus. This can precipitate acute pulmonary oedema, especially in patients with stenotic valvular lesions. Thus, little fluid is administered during delivery and caesarean sections (100-200ml at a time), and a degree of postpartum haemorrhage (PPH) is tolerated depending on the state of right ventricular filling on repeated echocardiograms. With minimal fluid administration, the autotransfusion of 400-500mL of blood is often nicely balanced by 400-500mL of bleeding to maintain a state of compensated normovolaemia.

Arrhythmias

Parturients will often already be started on anti-arrhythmic therapies, which can be loaded again in the peripartum period. Modification of the ALS algorithm for the pregnant patient (manual uterine displacement, perimortem LUSCS) remains unchanged, and there is a low threshold for cardioversion for any unstable rhythm.

Oxytocics and postpartum haemorrhage

The haemodynamic consequences of postpartum haemorrhage can be significant, but oxytocics themselves carry substantial risks, particularly in patients with mitral stenosis. Ergometrine increases cardiac afterload through vasoconstriction, increases pulmonary arterial pressures (PAP) and can lead to coronary vasospasm.[44] Carboprost can also lead to increases in PAP, and they are both avoided.[45] In patients with pre-existing pulmonary hypertension, a sudden increase in PAP can precipitate a pulmonary hypertensive crisis and acute right heart failure. Oxytocin causes hypotension and tachycardia through vasodilatation and has been associated with coronary vasospasm. This combination of effects is particularly deleterious in patients with stenotic lesions, where diastolic filling is paramount in maintaining cardiac output. Therefore, oxytocin must be carefully titrated in 0.5 to 1 unit doses and balanced with vasopressors based on real-time arterial monitoring. If required, a low volume infusion can be commenced.[46] Carbetocin carries similar risks to oxytocin but is less titratable and, therefore, omitted.[47] Misoprostol can be given rectally or orally because it has no cardiac side effects but is a weaker oxytocic.

Tranexamic acid is given prophylactically at the time of delivery to mitigate the risk of PPH. In the event of significant bleeding, early surgical management is required and haemostasis can often be achieved in the setting of a well-working epidural. Minor suturing or placement of a Bakri balloon can be performed quickly in intensive care, whereas other procedures may require transfer to the operating theatre.

Obstetric emergencies and other drugs

Respect for a fragile haemodynamic state should be observed with any drug given during the peripartum period. Seemingly benign drugs can have significant haemodynamic effects. This includes drugs used in the management of obstetric emergencies, such as terbutaline, which can cause tachycardia. There should be a clearly documented plan for how to manage such situations should they occur. Other anecdotal examples include significant hypotension with metoclopramide and nausea-related tachycardia from systemic absorption of neuraxial opioids.

Post-delivery management

Patients remain at high risk of decompensation and should be observed in a high dependency area with invasive lines left in situ in case of a return to theatre.[48] In addition to the risk of postpartum haemorrhage, there is a high risk of decompensated cardiac failure with a decrease in CO, an increase in SVR, and a significant shift in fluid balance. Postpartum pain can also lead to sympathetic activation, and the epidural can be left in situ for analgesia if anticoagulation is not required. Intrathecal and epidural morphine can minimise the need for ongoing epidural infusions postoperatively. Breastfeeding should also be observed in the initial postpartum period as endogenous oxytocin release can have haemodynamic effects. Cardiology review and planning for potential surgery happen in the following weeks. Where possible, surgery is deferred for six months to allow for breastfeeding. This delay is often possible in the context of a return to a non-pregnant circulation and a reduction in transmitral pressures.

Anaesthetic techniques

Preoperative consultation with an anaesthetist should occur as early as possible. Unfortunately, this is often challenging due to the constraints of distance and also social and cultural barriers, which may not be readily apparent. Anaesthetic techniques are discussed with the patient and their family, in the presence of interpreters and Indigenous Liaison Officers, when required. Opportunistic review of patients is often necessary to align multiple assessments during a single visit (antenatal clinic, cardiology clinic, pre-anaesthetic clinic). Ideally, patients and their families have the opportunity to meet their multidisciplinary team well before delivery to build rapport and trust, and to ensure adequate time for questioning and understanding.

Labour in the intensive care unit

An early and well-organised arrival to ICU can ensure prompt insertion of invasive lines and an epidural and help facilitate early induction. This will maximise the chances of delivery during regular operating hours when staff and resources are optimised.

Arterial lines are placed for all patients and central lines for patients where vasoactive medications may be required (severe mitral stenosis, pulmonary hypertension, decompensated heart failure, arrhythmia). This is done before the insertion of the epidural to allow careful loading and titration. It also facilitates prompt emergency management during labour should a trip to the operating theatre be required. In our patient population, a small amount of sedation is frequently required to offset the tachycardia and sympathetic response from discomfort and anxiety associated with an unfamiliar environment and uncomfortable procedures. This can be in the form of remifentanil (0.1-0.2mcg/kg/min, similar to an obstetric PCA dose), which is easily titratable, reversible, safe and has favourable haemodynamic effects and small doses of midazolam (0.025mg/kg) once continuous CTG is placed.[49-51] Care must be taken to minimise the time in Trendelenburg position for central line insertion to prevent a sustained increase in venous return and acute pulmonary oedema.

An epidural is placed as normal to ablate the sympathetic response to labour. It is tested with low-dose local anaesthetic (5mL of 0.125% bupivacaine or 0.2% ropivacaine in 1mL increments). Further doses are given via a programmed intermittent epidural bolus or a patient-controlled epidural analgesia (PIEB/PCEA) regime utilising ultra-low dose concentrations of local anaesthetics to minimise motor block and haemodynamic effects (0.0625% bupivacaine or 0.1% ropivacaine). We use a PIEB with PCEA regimen as it has been shown to have a lower incidence of breakthrough pain, lower overall local anaesthetic dosing and minimal haemodynamic effects in the dilute concentration compared to continuous epidural infusion techniques.[52,53] Any vasodilation is counteracted by vasopressor support with real-time monitoring. Beta-blockade may be acutely necessary to curtail the reflex tachycardia associated with vasodilation from the epidural (or, in the case of a failed epidural). Both intravenous esmolol and metoprolol are safe and effective agents.

Artificial rupture of membranes (ARM) and induction are then performed with a reduced dose of oxytocin, followed by an intrapartum echo. Labour proceeds in the dependent position and passive descent is allowed at full dilation to minimise the undesirable effects of pushing. Instrumental lift-out is used with limited pushing if haemodynamics are favourable. Delivery ideally occurs in the presence of an anaesthetist and intensivist, with theatre notified in case of emergent transfer. While attempts are made to have a dedicated anaesthetic team available, the unpredictable nature of obstetrics and a small anaesthetic department dictates that all of our perioperative staff are prepared to manage these patients.

Management of the third stage and PPH is described previously above.

Caesarean section

Elective caesarean deliveries are performed with a slowly loaded epidural or a combined spinal epidural (CSE) technique. Where CSE is performed for high-risk patients, the spinal component contains only intrathecal morphine and no local anaesthetic. Intrathecal morphine is used to prevent postpartum sympathetic activation from incisional pain. The neuraxial technique is performed after invasive line insertion with or without sedation as described above. For patients with pulmonary hypertension, a pre-operative dose of nebulised milrinone 5mg can be given to vasodilate the pulmonary circulation. Epidural loading can be achieved with incremental 0.5% bupivacaine or 2% lignocaine with adrenaline 1:200,000, 1-2mL at a time, coinciding with slow increases in vasopressor support to offset any drop in SVR. For patients with pulmonary hypertension, vasopressin may be preferred to noradrenaline as a first-line option as it has no effect on pulmonary vascular resistance.

A semi-emergent LUSCS is possible with a titrated load of the epidural together with invasive monitoring and vasopressor support where time allows.

Elective caesarean sections under a general anaesthetic are recommended in cases where anticoagulation cannot be stopped, or the patient needs an emergent delivery (for maternal decompensation or foetal distress), where a slowly titrated epidural is not feasible.

While opioids are not globally utilised in GA LUSCS, in the cardiac patient, haemodynamic stability is prioritised over foetal sedation at induction. The use of remifentanil or alfentanil may provide the best balance between sympatholysis and prolonged foetal sedation. A general anaesthetic has its own risk, but it allows the use of a TOE and the delivery of inhaled nitric oxide, milrinone or nebulised prostacyclin in an intubated and ventilated patient.[41]

CONCLUSION

The perioperative management of rheumatic heart disease in the Top End falls upon the shoulders of a small group of dedicated and experienced obstetricians, midwives, cardiologists, intensivists, and anaesthetists. The management of this condition requires a comprehensive understanding of obstetric physiology, cardiac pathophysiology, and cultural awareness. As a result of ongoing social inequity we are likely to continue to encounter a high level of complicated obstetric RHD in the future. As a general tertiary hospital without cardiac surgical services, we continue to learn from our patients, colleagues, and experiences to improve and develop a service that fits the complex needs of patients in the Top End.

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