Management of congenital heart disease in the peripartum period: An illustrative case series

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ABSTRACT

More women with complex congenital heart disease (CHD) reach adulthood resulting in a cohort of patients who are at high risk for adverse events during pregnancy. The haemodynamic changes usual to pregnancy may be poorly tolerated in patients with poor systemic ventricular function, cyanosis, left-sided obstructive lesions and pulmonary hypertension. Complex CHD patients are best managed by a multi-disciplinary team at a high-risk centre. Pre-conception counselling aims at risk stratifying by means of a clinical evaluation, electrocardiogram and echocardiography. Echocardiography plays a vital role in delineating the initial lesions and residual lesions with its haemodynamic complications. The modified WHO (mWHO) classification provides a helpful tool to stratify anatomical and physiological lesions by maternal and foetal event rates and is recommended by the European Society of Cardiology (ESC). Patients with cyanosis, severe aortopathy and severe pulmonary hypertension fall into Class IV and termination of pregnancy is advised.

Patients may however choose to continue their pregnancy. We present 3 such cases of complex CHD (Fontan circulation, severe aortopathy and severe pulmonary hypertension) and illustrate some pertinent management principles in the peripartum period.

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INTRODUCTION

With the development of more advanced surgical techniques and better ICU care for the correction and palliation of congenital heart disease (CHD) in childhood, more than 90% of patients are now expected to survive to adulthood.[1] More women with moderate and complex CHD will therefore reach childbearing age and may desire pregnancy. Though mortality levels in high income countries (HICs) are low, there remains a considerable burden of morbidity during pregnancy particularly for those with complex disease.[2,3]

The outcomes of pregnancy depend not only on the complexity of the initial lesion, but the severity of residual lesions, systemic ventricular function, the presence of cyanosis, functional capacity (NYHA class) and pulmonary pressures.

HAEMODYNAMIC CHANGES IN PREGNANCY

The haemodynamic changes of pregnancy commence as early as the fifth week of gestation and peak during the second and third trimesters. Plasma volume expands disproportionately due to the increase in erythrocyte mass resulting in a dilutional anaemia. Cardiac output (CO) increases by 30% - 50%, due to an increase in blood volume, consequent stroke volume and later an increase in heart rate.[4] Both the systemic vascular resistance (SVR) and pulmonary vascular resistance (PVR) are reduced. These changes may unmask a structural lesion or exacerbate an existing CHD lesion. Poorly tolerated lesions include poor systemic ventricular function, severe obstructive lesions on the systemic outflow side, cyanosis and pulmonary hypertension.

During labour, uterine contractions and increased sympathetic activation due to pain result in a further 20% increase in CO. During the immediate post-partum period the risk of developing pulmonary oedema and other maternal complications remain high. Caval decompression, autotransfusion from the emptied and contracted uterus and the removal of the placenta produce further increases in preload. These changes usually revert back to normal within the 2 - 8 weeks post-partum.[4]

Pregnancy is a hypercoagulable state and poses an increased risk of systemic venous thromboembolism.

PRE-CONCEPTION COUNSELLING AND RISK STRATIFICATION

Ideally, all women with CHD are to be evaluated clinically, with an electrocardiogram (ECG) and transthoracic echocardiogram (TTE) prior to conception by a multi-disciplinary team to allow for adequate risk stratification, the setting of pregnancy expectations, anticipation of complications and its management to ensure successful outcomes as best possible.[5] The most common risks to the mother include arrythmias, heart failure, thromboembolic complications and pre-eclampsia. Frequent adverse risks to the foetus include premature birth, small for gestational age (SGA), neonatal death and recurrence of CHD.

The initial step is to evaluate the patient clinically, paying particular attention to symptoms. The NYHA class may be difficult to determine when already pregnant since symptoms of fatigue, breathlessness, palpitations and oedema may be inherent to pregnancy as well. ECG and TTE allow for the assessment of residual lesions, its presence and anatomical and physiological severity. Patients are then risk stratified according to the modified World Heart Organisation (mWHO) classification.[6] Class I includes mild disease that results in only a small increase in morbidity and no increase in mortality to the mother when compared to the general population, e.g. mild pulmonary stenosis or a repaired atrial septal defect (ASD). Class IV poses the highest risk of mortality and morbidity to both mother and foetus. Unless adequate and easily accessible clinical, surgical and obstetric expertise exist in your institution, those in Class IV are advised to terminate the pregnancy. Continuing the pregnancy would pose significant risk of mortality to the mother. Surgical correction prior to conception is advised and alternatives to pregnancy discussed. This is a sensitive issue and embarked on respectfully with the relevant family members.

The CARPREG I[7] and II[8] and ZAHARA[9] scores are alternative risk stratification models to mWHO, but it is advised that one scoring system is used consistently in your region amongst attending clinicians.

ECHOCARDIOGRAPHY IN PREGNANCY

TTE remains the most accessible and cost-effective modality to assess anatomy and physiology during pregnancy. It is indicated in pregnant women with unexplained or new cardiovascular signs or symptoms. When reporting, it is important to note both the gestational age and position of the patient (supine vs. left lateral decubitus) as the position of the gravid uterus may result in a change in preload or afterload.

All cardiac chambers dilate during pregnancy due to the increase in blood volume – the timing and degree of dilatation being variable. The chambers usually remain within the upper limit of normal though. Other changes include an increase in LV wall thickness and LV mass.[10]

Pregnancy does not change the pre-existing valve stenosis severity but transvalvular velocities are increased due to increased blood flow that occurs during pregnancy.

Velocity-derived pressure gradients therefore correlate less well with pre-pregnancy gradients. Measuring trans-valvular flow with doppler remains useful as it can be used to calculate valve area and stenosis severity even in presence of the increased blood flow. Mild increases in degrees of valve regurgitation occurs during pregnancy, resulting from an exaggerated separation of leaflets due to chamber dilatation, mainly affecting the mitral, tricuspid and pulmonary valves.[10]

Parameters that remain largely unchanged during pregnancy include ejection fraction, fractional shortening, E/e' ratio and RVSP. Some debate exists on whether LV systolic function remains normal or increases. Trace to mild pericardial effusions may be present.[11]

Trans-oesophageal echo (TOE) can be done relatively safely but with an increased risk of vomiting/aspiration. It is reserved for cases where TTE assessment is inadequate. Cardiac Magnetic Resonance imaging (CMR) is reserved for selected indications only.[11]

CASE 1: FONTAN CIRCULATION

A 29-year-old patient with a hypoplastic RV, double inlet LV underwent a fenestrated total cavo-pulmonary circulation (TCPC) at age 9. The fenestration was subsequently closed. She reports no effort intolerance prior to her current pregnancy and has a baseline saturation of 86% at rest. She defaults appointments in between pregnancies and has disregarded all pregnancy and contraception-related advice. She has had 2 previous pregnancies: A mid-gestation miscarriage

and a "successful pregnancy" in 2004 when she presented at 32 weeks gestational age in labour. She now presents having delivered a baby at home at 29 weeks gestation. This was an unbooked and unplanned pregnancy.

On examination, she was cyanosed with oxygen saturation of 72% on room air (Figure 1 A). She was lethargic and dehydrated, having suffered profound blood loss with active ongoing post-partum haemorrhage. Her heart rate was 138bpm and BP92/52. Her chest was clear and JVP not elevated. She has a single second heart sound with a flow murmur at the second inter-costal space.

TTE showed a hypoplastic RV and double inlet LV. No significant AV valve regurgitation was noted. The inferior limb of the Fontan circuit was seen with normal pressures. No fenestration was seen (Figure 1 B - D).

She was treated with a blood transfusion and rehydrated. Retained products were removed. Saturations improved to baseline. A progesterone-based contraceptive was implanted before discharge.

Even patients with an optimally functioning Fontan circuit, remains at increased risk of maternal and foetal complications (mWHO III). As was the case with this patient, those with cyanosis, poor ventricular function, moderate to severe atrio-ventricular valve regurgitation (AVVR) and protein-losing enteropathy (PLE) should be counselled against pregnancy (mWHO IV) citing the high risk of foetal loss and maternal mortality.[6] It remains important to stress that despite choosing pregnancy against medical advice, the pregnancy risks are still best managed in a high risk clinic and patients encouraged to do so.

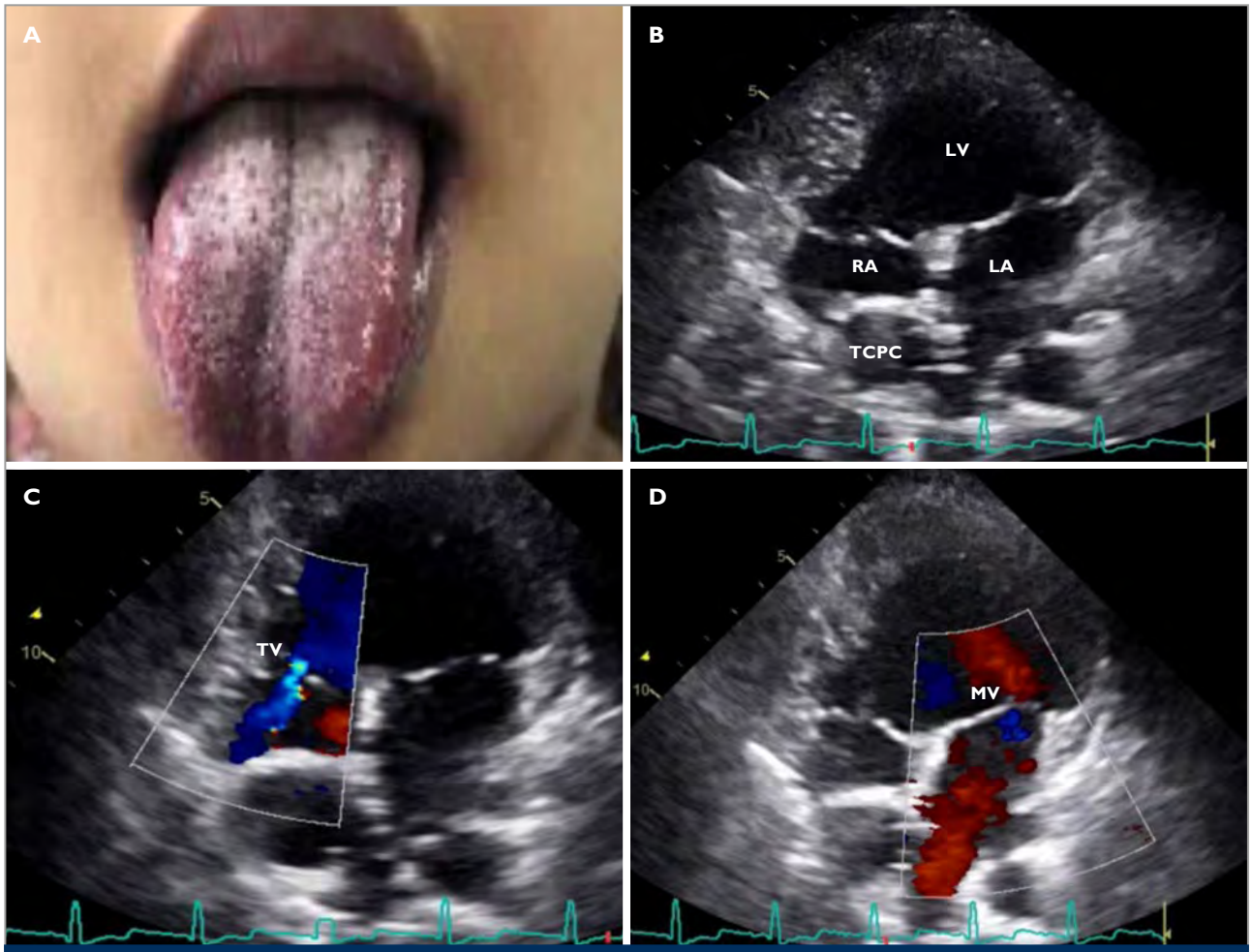

FIGURE 1: A 29-year-old patient with a total cavo-pulmonary circulation (TCPC) presented on day one post-partum with marked dehydration and cyanosis (A) *(Taken with patient permission)*. Apical view shows a double-inlet LV and the TCPC conduit (B). The pressures in the TCPC conduit were low. C and D showing no significant tricuspid- or mitral incompetence.

A recent study of maternal and foetal outcomes in Fontan patients showed that despite the recommendation, only a small number of patients received pre-conception counselling (8%). No maternal deaths occurred over a 12-year follow-up period. Heart failure and arrhythmias were the most frequently encountered maternal complications. All women with baseline oxygen saturations of <85% miscarried and 51% of those with saturations >85% miscarried. The rate of miscarriage is higher in Fontan circulation compared to other forms of CHD. Over 70% of patients delivered before 37 weeks gestation.[12]

Women with a Fontan circulation have a greater risk of post-partum haemorrhage compared to patients with other cardiac lesions. Post-partum haemorrhage is poorly tolerated in Fontan patients as optimal functioning of the circuit is preload dependent.[13] Blood loss is to be kept to a minimum and hydration maintained. Use oxytocin with caution, as this may result in systemic vasodilatation and pulmonary vasoconstriction. In patients with concomitant systemic ventricular dysfunction and AV valve regurgitation, be cautious to not fluid overload them, as balancing adequate diuresis while maintaining an adequate preload may be challenging.

Both the Fontan circulation and pregnancy are prothrombotic states but the preferred strategy for anticoagulation remains controversial and no consensus guidelines currently exists. Each patient is to be risk stratified on a case-by-case basis.

CASE 2: HIV-ASSOCIATED AORTOPATHY

A 31-year-old unbooked patient presented at 35 weeks gestation in heart failure and in early labour. Her past history includes aortic incompetence and a dilated aortic root diagnosed incidentally at the time of presenting with pulmonary tuberculosis in 2010. She declined HIV testing at the time, but tested positive in 2014 with a recurrence of pulmonary tuberculosis. Anti-retroviral therapy was commenced, and she was offered aortic root and valve surgery. She declined surgery at the time and defaulted treatment and follow-up.

TTE showed a markedly dilated aortic root with severe aortic incompetence (Figure 2 A - C). The LV was dilated with preserved systolic function. Due to the emergent nature of her being in labour, an MRI was not performed in this case. A caesarean section was done under regional anaesthesia with both vascular and cardiothoracic surgical teams on stand-by to perform emergency aortic surgery if required. Despite extensive counselling to her and her family, the patient regrettably once again refused aortic root surgery after caesarean section.

Aortopathies are a well-recognised cause of maternal death in pregnancy.[14] Oestrogen has been shown to inhibit collagen and elastin deposition in the aorta but it remains debated

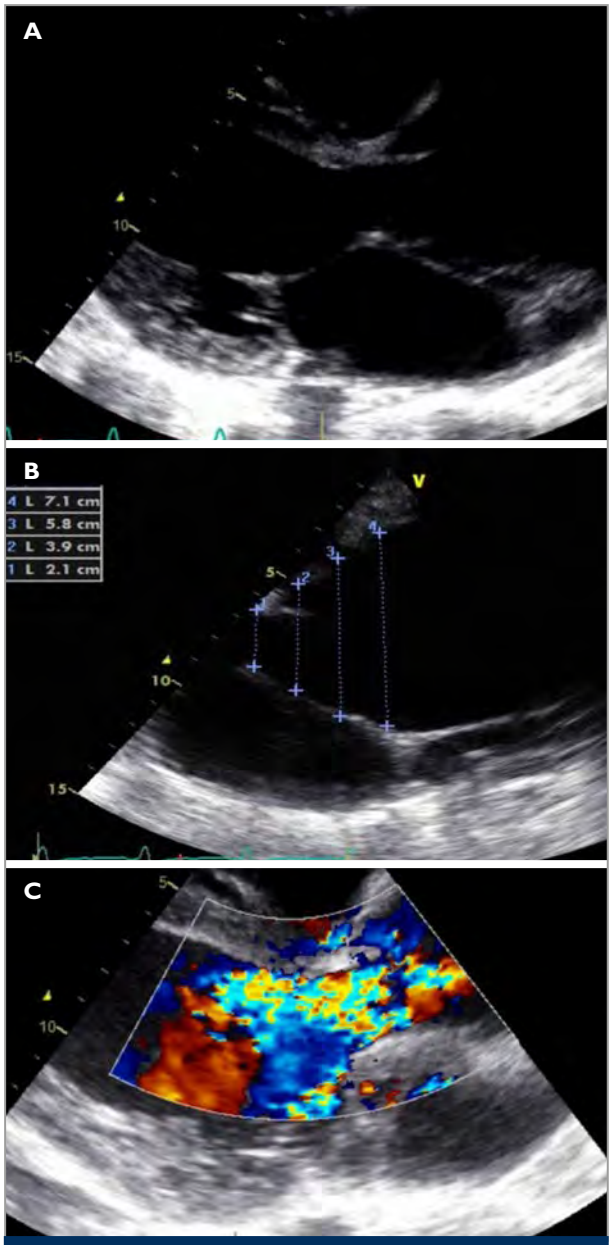

FIGURE 2: A 31-year-old unbooked patient presented at 35 weeks gestation in heart failure. (A) A parasternal long axis view depicting a markedly dilated aortic root. (B) Aortic dimensions showed an aortic annulus dimension of 2.1cm , sinus diameter 3.9cm, sino-tubular junction 5.8cm and ascending aortic diameter 7.1cm. (C) Parasternal long axis with colour flow doppler shows severe aortic incompetence.

whether pregnancy worsens aortic dilatation. The mortality rate from aortic dissection in pregnancy and early post-partum remains high with the major determinant of risk being aortic root size.

The mWHO criteria currently risk stratifies patients with aortopathy as follows:[6]

- If no aortic root dilatation - Category II
- Aortic root diameter 40 - 45mm – Category III
- Aortic root diameter >45mm – Category IV (highest maternal mortality risk and therefore a contraindication for pregnancy)

TTE is the first-line imaging modality but since it does not delineate the extent of the aneurysm or the presence of a dissection, pan-aortic advanced imaging, preferably MRI without gadolinium is recommended. If not available, computed tomography (CT) would be indicated. Monthly or bi-monthly surveillance imaging of the aortic root is advised using TTE.[6] Betablocker therapy is widely prescribed though no robust studies have shown a reduction in dissection rates with its use. With the exception of Atenolol, betablockers have a favourable safety profile but are associated with increased risk of small for gestational age (SGA), neonatal bradycardia and hypoglycaemia.[15]

Delivery by caesarean section is advised in patient with an aortic root diameter >45mm to avoid the strain of contractions and reduce Valsalva manoeuvres.

The true incidence and prevalence of HIV-associated aneurysms is unknown. A South African study reported an incidence of symptomatic HIV vasculitis of 1%.[16] There are no specific recommendations for the management of HIV-associated aortopathy and standard treatment principles are followed. In symptomatic patients (aortic regurgitation with heart failure symptoms, dissection or rupture) emergent surgery is indicated. If asymptomatic, but aortic root diameter >55mm, the risk of dissection and rupture is high and surgery is recommended.

CASE 3: SEVERE PULMONARY HYPERTENSION

A 27-year-old patient presented with dyspnoea and pre-syncope at 15 weeks gestation. She has a prior diagnosis of atrial septal defect (ASD) but declined surgery previously. On clinical examination she was not clubbed or cyanosed with saturations of 94% on room air. She had a prominent left precordial chest wall bulge, a BP of 105/62 and fixed splitting of S2 with a loud P2 component. No distinct murmurs were heard.

Her CXR (Figure 3A) showed a normal cardiothoracic ratio, large pulmonary arteries and plethoric lung fields. Right ventricular hypertrophy (RVH) with strain was noted on ECG. On TTE, a secundum ASD with a dilated RV and RVH was seen. Colour flow doppler showed a left to right shunt (Figure 3 B and 3 C) An RVSP of 82mmHg was measured. On right heart catheterisation, pulmonary pressures were 89/34 with a mean of 53mmHg (>2/3 systemic pressures) (Figure 3 D). A step up of saturations was noted in the right atrium. A Qp:Qs of 2.78:1 and PVR of 4.5 was calculated. No bidirectional shunting was noted. Wedge-to-free pullback showed a marked diastolic gradient signifying pre-capillary pulmonary hypertension, in the presence of a normal wedge mean pressure. We advised termination of pregnancy but upon consultation with her family, the patient decided to against this. Admission to the obstetrics unit and the commencement of Sildenafil was advised. She moved provinces, continued the Sildenafil and went into spontaneous labour at 32 weeks gestation. An epidural was administered, resulting in an acute drop in BP. She developed an acute pul-monary crisis and demised.

Pregnancy in women with pulmonary hypertension portends a high risk of maternal mortality (30% - 50%), regardless of aetiology, even with optimal medical management. Most deaths occur in the third trimester or post-partum period, mainly due to increases in PVR resulting in pulmonary hypertensive crises. Heart failure and arrhythmias are frequent causes of maternal morbidity.[17] Foetal complications include premature delivery (64%), SGA (37.5%), and foetal mortality (9.5%).[1] The mWHO score therefore categorises it as Class IV, contra-indicates pregnancy and recommends first trimester termination of pregnancy.

Although ASDs are well tolerated in pregnancy, the presence of marked RV dilatation, RV dysfunction and severe PHT is poorly tolerated. The PVR and SVR falls to the same degree and shunt size does not usually change. If significant pulmonary vascular disease is present, the usual pregnancy-related fall in PVR is diminished. Furthermore, the drop in SVR during pregnancy, when pulmonary hypertension is already present, can result in or exacerbate a right to left shunt.

For those who continue the pregnancy, there are no standardised protocol for management of pulmonary hypertension in pregnancy. They should be managed by a high-risk centre and multi-disciplinary team. TTE plays an important role in defining the diagnosis, the degree of severity and the aetiology of pulmonary hypertension. It may overestimate pulmonary artery pressures (RVSP surrogate) in pregnancy, when compared to cardiac catheterisation – up to 32% of pregnant patients are misclassified as pulmonary hypertension (PAH) on echo when they had normal pressures on cardiac catheterisation.[18]

Patients are advised to avoid high altitude travel. Hospitalisation from 20 weeks gestation for bedrest is advised. Anticoagulation is recommended.[19] The main objectives of treatment are to optimise RV preload and systolic function, reduce the PVR, and prevent ischemia to maintain RCA coronary perfusion pressure. Diuretics may be needed to reduce volume overload must be used with extreme caution to prevent over-diuresis and a drop in preload.

Pharmacological management with epoprostenol has been shown to improve exercise capacity, quality of life, haemodynamic and long-term survival in pulmonary arterial hypertension. Endothelin receptor antagonists and riociguat are contraindicated. Tresprostinil and inhaled iloprost has not been studied in pregnancy and is to be avoided. In high-income countries, patients on phosphodiesterase 5 inhibitors and bosentan may be converted to IV epoprostenol, administered via hickman lines. Oral sildenafil remains the therapy of choice in many countries. Given the risks of sudden systemic vasodilation, it is to be commenced slowly, and in hospital.

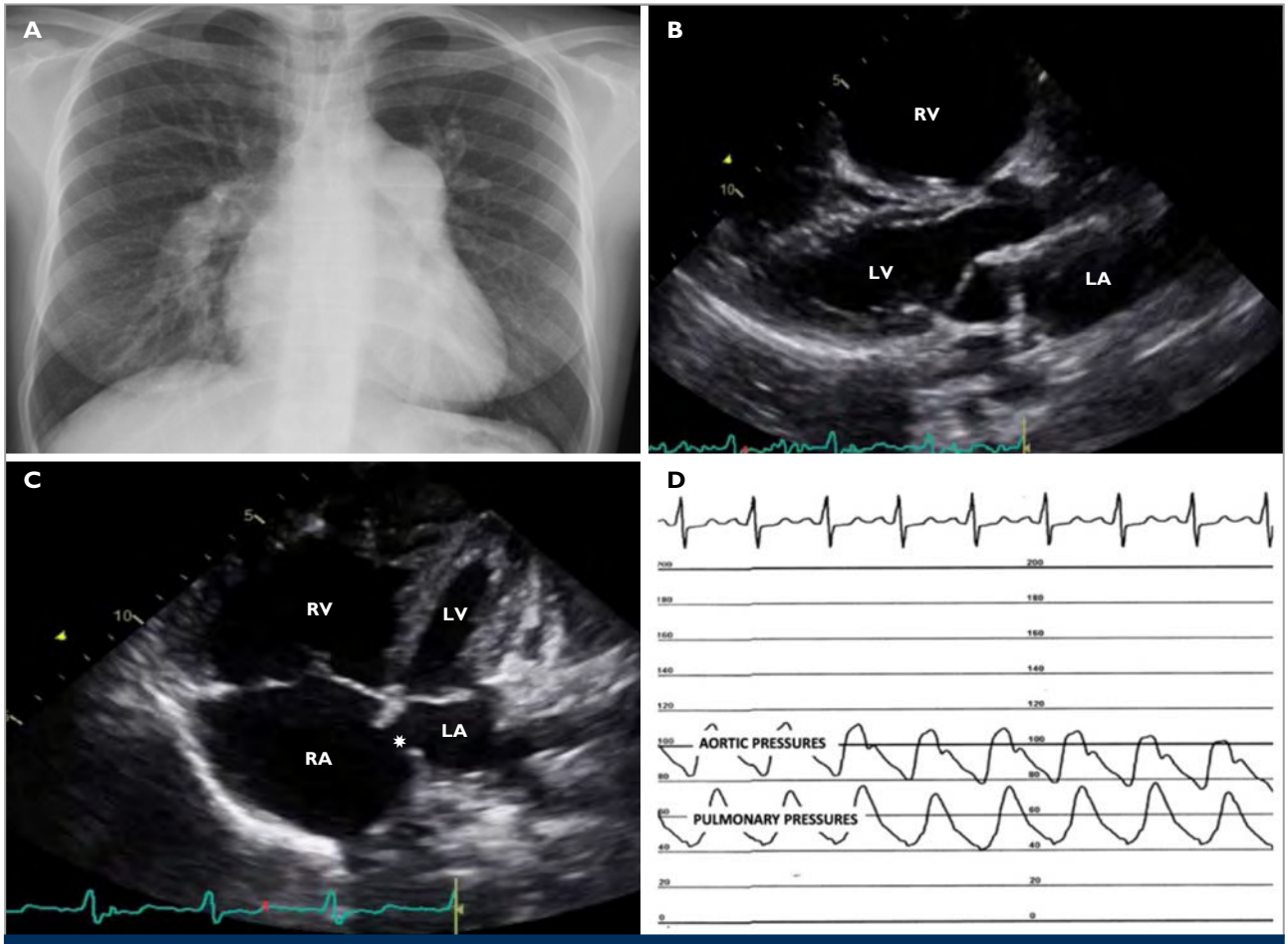

FIGURE 3: A 27-year-old patient, known with an atrial septal defect (ASD) presented with dyspnoea and pre-syncope at 15 weeks gestation. (A) Chest x-ray showed a normal cardio-thoracic ratio, markedly enlarged pulmonary arteries and plethoric lung fields. (B) A parasternal long axis shows a markedly dilated RV outflow tract and a pressure loaded RV. (C) The apical 4-chamber view shows a dilated RV and RA with a secundum ASD (white star). At cardiac catheterisation, severe pulmonary hypertension was noted. (D) The systolic pulmonary pressures were >2/3 of the systolic pressure.

The timing and mode of delivery, detailed in advance by the multi-disciplinary team, aims at an early planned delivery at 32 - 36 weeks gestation. There are conflicting studies regarding the optimal mode of delivery. Vaginal delivery is thought to result in less risk bleeding, infection, and venous thrombo-embolism. Avoid a prolonged second stage of labour as valsalva decreases venous return resulting in hypotension since they are pre-load dependent. Most guidelines recommend caesarean section with regional anaesthesia (avoid a general anaesthetic).[20] Be careful not to administer a single dose spinal anaesthesia to reduce the risk of sudden hypotension and the induction of a pulmonary hypertensive crisis. Close cardiac monitoring for up to 2 weeks post-partum is recommended since complications occur mostly at term or in the immediate post-partum period.

CONCLUSION

Complex CHD patients are to be managed in high-risk centres by a multidisciplinary team. Risk stratification is best done pre-conception. Echocardiography plays and important role in the follow-up during all stages of pregnancy as well as guiding delivery and emergent situations. It is important to know pre-pregnancy lesions, residual lesions and the anticipated effects of physiological change on the lesions to develop safe and effective pregnancy care-plans. These plans are to be effectively communicated to all relevant staff (including the on-call team).

Conflict of interest: none declared.

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