Cardiovascular, Obstetric and Neonatal Outcomes in Women with a Previous Fontan Repair

Samantha Bonner[1], Omar Asghar[2], Anna Roberts[1], Sarah Vause[1], Bernard Clarke[2], Bernard Keavney[2]

[1]*St Mary's Hospital, Central Manchester NHS Foundation Trust, UK*

[2]*Manchester Heart Centre, Central Manchester NHS Foundation Trust, UK*

Background

The Fontan surgical procedure has significantly improved prognosis and survival for patients with complex congenital heart disease characterised by univentricular physiology. Consequently, an increasing number of post Fontan repair patients reach childbearing age and contemplate pregnancy. Pregnancy in post Fontan repair patients is associated with an increased risk of cardiovascular, obstetric and neonatal complications. This increased risk may be attributable to the cardiovascular changes associated with pregnancy; primarily volume loading and a hypercoagulable state. In order to enable post Fontan repair women to make informed decisions regarding family planning and pregnancy, it is vital that clinicians provide them with reliable and contemporary information.

Objectives

To determine cardiovascular, obstetric and neonatal outcomes of pregnancies in women who have a Fontan circulation.

Methods

A retrospective case note review of all women with a Fontan circulation who attended the joint obstetric cardiac antenatal clinic at St Mary`s Hospital, Manchester (UK) between 2004-2016 was performed.

Results

In total, there were 19 pregnancies in 9 women with a history of Fontan repair. 10 pregnancies (53%) resulted in live birth; 1 in a stillbirth at 31 weeks gestation and 8 in miscarriages. Cardiovascular complications occurred in 2 pregnancies (11%). There were no thrombotic events, arrhythmias, myocardial infarction or endocarditis in the antenatal or postnatal period. Obstetric complications included miscarriage (26% first trimester, 16% second trimester), along with premature delivery (24-36+6 gestational weeks)(80%) and fetal growth restriction (70%). Just over half of the women were delivered by caesarean section (60%).

Conclusions

Pregnancy loss, pre term birth and fetal growth restriction are all more common in women with a Fontan repair. Multidisciplinary, pre conceptual assessment and counselling are important to ensure women make informed decisions regarding future pregnancy.

Clinical Outcomes of Women with Peripartum Cardiomyopathy with and without Preeclampsia

Isabelle Malhame, Natalie Dayan, Cristiano Soares De Moura, Evelyne Vinet, Louise Pilote

Background

Preeclampsia (PE) is a potent risk factor for peripartum cardiomyopathy (PPCM), a condition associated with significant maternal morbidity. The extent to which PE affects PPCM-associated outcomes requires further study.

Objective

To evaluate whether women with PPCM and PE have different outcomes compared with women with PPCM without PE.

Methods

We conducted a retrospective cohort study using the *MarketScan Commercial Claim and Encounters Database.* We included all women with a delivery between April 1st 2011 and June 30th 2014 identified through ICD-9-CM diagnoses and procedures. We excluded women with cardiac conditions at estimated date of conception. We identified women who had PPCM with PE (pePPCM group) and PPCM without PE (npePPCM group). We compared the proportions with Major Adverse Cardiac Events (MACE), Severe Cardiovascular Maternal Morbidity (SCMM) and death between groups using Chi-square or Fisher's exact test. We calculated crude risk ratios with their 95% confidence intervals for MACE, SCMM and death. Estimates were adjusted for age, obesity, multiple pregnancy, diabetes, and hypertension.

Results

We identified 286 women with pePPCM, and 533 women with npePPCM. Women with pePPCM were more likely to be obese, have chronic hypertension, gestational diabetes and chronic kidney disease and more likely to have multiple gestations. Women with pePPCM were more likely to present with acute heart failure (21.7% versus 16.1%), pulmonary oedema (27.3% versus 14.6%), and respiratory distress (21.7% versus 15.2%). Accordingly, pePPCM patients were at higher risk of MACE (42.7% versus 31.5%; adjusted RR 1.41 95% CI (1.01, 1.95)) and SCMM (53.8% versus 37.5%; adjusted RR 1.71 95% CI (1.24, 2.35)). However, we found that the risk of death might be lower in women with pePPCM (0.3% versus 1.1%; crude RR 0.31 (0.02, 1.82)).

Conclusion

In our cohort, women with pePPCM had a higher risk of MACE and SCMM suggesting a different disease mechanism.

Pregnancy in Women with a Fontan circulation - is there an effect on maternal outcomes post partum?

Emily Moroney[1], Diana Zannino[1], Yves d'Udekem[1,2,3], **Dominica Zentner**[2,4]
[1]*Murdoch Children's Research Institute, Australia*
[2]*University of Melbourne, Australia*
[3]*Royal Chidren's Hospital, Australia*
[4]*Royal Melbourne Hospital, Australia*

Background

Pregnancy in the woman with a Fontan circulation is categorized as moderate to high risk of maternal morbidity and mortality. However, there are no reports of maternal mortality during pregnancy. Longer-term outcomes, including survival, which may be adversely affected by pregnancy in this population, remain unknown.

Objective

To compare outcomes in women with a Fontan circulation that have and have not had a pregnancy, within the population of the Australia and New Zealand (ANZ) Fontan Registry.

Methods

The ANZ Fontan Registry currently has 1,466 individuals enrolled, of whom 265 are women (surviving beyond 18 years of age with adult cardiology follow up data) (at last data update, end 2015) Of these, 30 women report 46 pregnancies beyond 20 weeks gestation, resulting in 43 live-births, (1 set of twins, 4 stillbirths). In the other 233 women, a small number of miscarriages are reported (n = 10) (Note: 2 women declined to share pregnancy information)

Results

The women who have had a pregnancy are older 32.9 (26.6-37.3) years compared with 25 (20.5-29.3) years, and appear to have had less pre pregnancy medical events reported (Figure 1). This would suggest either appropriate pre pregnancy assessment and advice, or possibly an increased miscarriage rate in women who are less well. Post-partum follow up was available for 26/30 women, with a median time since first pregnancy of 3.6 (1.2-7.5) years. There appear to be an increase in thromboembolic events post pregnancy. This data does not support a difference in mortality, at this limited post-partum follow-up.

Conclusion

The importance of long term follow up of these women is highlighted by this data. Additionally, the need exists to match women for significant factors such as age, type of Fontan and systemic ventricle morphology in order to ensure that the interpretation is accurate.

The Role of Cardio-Pulmonary Exercise Testing in Predicting Outcomes in Mothers with Congenital Heart Disease

Panagiotis Mastrogiannopoulos[2], Daniel Mallaber[1], Angella Bryan[1]
[1]*Manchester University NHS Foundation Trust, UK*
[2]*University of Manchester, UK*

Background

An expanding population of women with congenital heart disease (WCHD) now reach childbearing age. This number continues to rise, particularly amongst those with complex ACHD. Cardio-Pulmonary Exercise Testing (CPET) variables have previously been linked to pregnancy outcomes in WCHD.

Objective

This study aimed to identify associations between pre-pregnancy CPET variables and pregnancy outcomes in a cohort of WCHD, and to compare CPET to existing CARPREG and WHO models for risk prediction.

Method

A single-centre retrospective observational study of WCHD who had undergone CPET up to 6 years prior to becoming pregnant was performed. WHO classification and CARPREG score were retrospectively ascertained.

Results

52 pregnancies (age at delivery, 29±5 years), in 37 WCHD were identified. Of these pregnancies 38 (73%) were successful, 10 (19.2%) miscarried before 16 weeks and 4 (7.8%) suffered a fetal death after 16 weeks. The mean time from most recent CPET to delivery was 2.7 (±1.7) years. Maternal cardiac, obstetric, and neonatal complications occurred in 6 (11.5%), 24 (61.5%) and 22 (52.4%) of pregnancies respectively. Pre-term delivery (2 at Anaerobic Threshold (p=0.001, OR (95% CI) = 1.66(1.21,2.27)). The association between WHO classification and pre-term delivery had wide confidence intervals. There was no association between CARPREG and pre-term delivery. Of the 13 pre-term deliveries 7 (53.8%) were planned and 6 were unplanned (46.2%). No associations were found between other outcomes and CPET, CARPREG or WHO classification.

Conclusion

CPET may outperform WHO and CARPREG in predicting certain pregnancy outcomes in WCHD. More large-scale studies are needed to confirm associations between CPET and outcomes, and to compare the sensitivity of CPET, CARPREG score, and WHO classification.

Short and Long Term Impact of Pregnancy on Women with a Systemic Right Ventricle

Kathryn Lindley[1], Shayna Conner[2], Alison Cahill[2], Stephanie Velloze[1], Joseph Billadello[1]
[1]*Washington University in St. Louis, USA*
[2]*Washington University in St. Louis, USA*

Background

Data regarding safety of pregnancy in women with a systemic right ventricle (RV) is limited.

Objective

To assess maternal and fetal outcomes and long-term RV function decline in women undergoing pregnancy with a systemic RV.

Methods

This retrospective cohort of patients delivering at Barnes-Jewish Hospital from 2003-2013 compared maternal and fetal outcomes in women with a systemic RV (D-transposition of the great arteries (D-TGA) with atrial switch or L-TGA) to patients with Tetralogy of Fallot (ToF), a congenital heart defect of comparable complexity but with a systemic left ventricle (LV). Long-term systemic RV function was assessed by comparing echocardiograms from baseline (pre-conceptual or first trimester) and at most recent follow up, and were compared to a control group of nulliparous women with systemic RV.

Results

Pregnancy outcomes were assessed in 21 women with systemic RV and 19 with ToF. Live birth occurred in 86% of ToF and 96% of TGA patients (remainder elective abortions). The incidence of pregnancy-related cardiac complications (heart failure, arrhythmia, thromboembolism, death) was comparable (9.1% Tof vs 4.5% TGA, p=0.6). No difference in obstetric or fetal complications was observed (obstetric: 27.3% ToF vs 22.7% TGA, p=0.7; fetal: 18.2% ToF vs 22.7% TGA, p=0.7). Long-term echocardiographic follow up (mean 68 +/- 35 months) was available in 20 pregnant and 13 nulliparous women with systemic RV. Baseline RV function was comparable between the two groups. At latest follow up, there was no significant difference in RV Tei (p=0.8), progressive RV dilation (p=0.16) or progressive global RV dysfunction (p=0.7) after adjusting for age, follow up time, and baseline measure.

Conclusion

The incidence of adverse cardiovascular outcomes in pregnancy in women with a systemic RV is low, and comparable to outcomes in ToF. Pregnancy does not seem to accelerate decline in RV function beyond the natural history of the disease.

Pregnancy Outcomes in Women with Tetralogy of Fallot:

Data from the European Society of Cardiology Registry on Pregnancy and Cardiac disease (ROPAC)

Lucia Baris[1], Roger Hall[2], Jolien W. Roos-Hesselink[1]
[1]*Erasmus Medical Center, Netherlands*
[2]*University of East Anglia and Norfolk, UK*

Background

The hemodynamical changes of pregnancy can be challenging in the case of underlying maternal heart disease. The outcomes of pregnancy in women with Tetralogy of Fallot are not intensively investigated.

Objectives

To describe the outcomes of pregnancy in women with Tetralogy of Fallot (ToF).

Methods

Within the international prospective Registry On Pregnancy And Cardiac disease (ROPAC), we describe cardiac, obstetric and fetal outcomes of pregnancy within patients with ToF and identify predictors of adverse cardiac outcome.

Results

In the 240 included ToF patients (mean age 28.7 ± 5.1 years) no maternal mortality occurred. In 18 pregnancies (8%) at least one cardiac event occurred, of which heart failure was the most common complication (n = 11, 5%). Ventricular tachyarrhythmia's complicated 7 pregnancies (3%) and supraventricular tachyarrhythmia's occurred in 2% (n = 5). One patient (0.5%) suffered from valvular prosthesis thrombosis. Seven patients (3%) had not undergone complete correction of ToF, of which 4 patients (2%) had only undergone either a surgical correction of VSD or valvular replacement. At least one obstetric event occurred in 10 patients (4%), with postpartum hemorrhage in 4 patients (2%) and pre-eclampsia in 3 (1%). There were 3 miscarriages (1%) and 4 patients suffered from pregnancy-induced hypertension (2%). Fetal events occurred in 40 patients (17%), with 3 cases of late fetal mortality (1%). Preterm birth (37 weeks) and low Apgar score occurred in 34 (14%) and 9 patients (4%) respectively. No predictors of adverse cardiac or obstetric events were found, while the use of beta blockers was associated with adverse fetal outcome.

Conclusion

Most women with ToF tolerate pregnancy well, and can safely embark upon pregnancy. However, cardiac and fetal complications were not uncommon. The use of beta blockers during pregnancy was associated with fetal complications.

Direct Oral Anticoagulants during Pregnancy or the Post-partum Period, Current Evidence in Humans

Heleen Lameijer[1,2], Petronella Pieper[1], Jan Aalbers[1], Karina Meijer[3], Dirkjan van Veldhuisen[1]

[1]*University Medical Centre Groningen, University of Groningen, Netherlands*
[2]*University Medical Centre Groningen, University of Groningen, Netherlands*
[3]*University Medical Centre Groningen, University of Groningen, Netherlands*

Introduction

Direct oral anticoagulants (DOACs) are increasingly used for anticoagulation or prevention of thromboembolic events in conditions that may co-occur with pregnancy, such as deep venous thrombosis, pulmonary embolism and atrial fibrillation. However, evidence regarding efficacy and safety during pregnancy is scarce.

Aim

To systematically review the current literature concerning the efficacy and safety and pregnancy outcome of DOACs during pregnancy in human.

Methods

Systematic review for studies published up to 04-07-2017.

Results

236 cases of DOAC use during pregnancy were reported in recent literature (2014-2016). Rivaroxaban was the most reported DOAC (n=178, 75%), followed by Dabigatran (n=27, 11%), Apixaban (n=21, 9%) and Edoxaban (n=10, 4%). DOACs were mostly used for prophylaxis or treatment of DVT (n=91, 94%). DOACs were discontinued within the first 2 months of pregnancy in 84%, with maximum reported duration of 26 weeks. Pregnancy outcome data were available for 59% of the pregnancies, of which 28% were electively terminated. In ongoing pregnancies total miscarriage rate was 31% and live birth rate was 68%. Fetal and neonatal abnormalities were reported in 8%, of which at least half (bone and facial structural abnormalities) are suspected to be related to Rivaroxaban use during the 1st trimester of pregnancy. Thrombotic and obstetrical complications were highly underreported (18% reported).

Conclusion

Safety and efficacy of the use of DOACs during pregnancy is not supported by current literature. The limited available evidence raises concern regarding embryo-foetal safety, with high incidence of miscarriages and at least a 4% rate of anomalies with the use of Rivaroxaban.

Long-term Cardiac Function after Peripartum Cardiomyopathy and Preeclampsia: a Danish Nationwide, Clinical Follow-up Study using Maximal Exercise Testing and Cardiac Magnetic Resonance Imaging

Anne Ersboell[1], Annemie Bojer[2], Maria Hauge[1], Marianne Johansen[1], Peter Damm[1,3], Finn Gustafsson[2], Niels Vejlstrup[2]

[1]*Rigshospitalet Copenhagen University Hospital, Denmark*

[2]*Rigshospitalet Copenhagen University Hospital, Denmark*

[3]*University of Copenhagen, Denmark*

Background

Recent studies have shown that most women with peripartum cardiomyopathy (PPCM) recover left ventricular ejection fraction (LVEF) within a year after diagnosis, but clinical studies of the long-term prognosis after PPCM are few.

Objective

To evaluate cardiac structure and function after PPCM in a nationwide cohort of women diagnosed in Denmark from 2005 to 2014.

Methods

58 women with PPCM were invited to participate in a clinical follow-up study including maximal cardiopulmonary exercise testing and cardiac magnetic resonance imaging. For comparison, two matched groups of women with previous severe preeclampsia and previous normal pregnancies, respectively, were invited too.

Results

A total of 28 women with PPCM accepted the invitation (48%) together with 28 women with previous preeclampsia and 28 women with previous normal pregnancies. Median time to follow-up was 91 months (range 27 – 137). The majority of women with PPCM had no symptoms of heart failure and recovered cardiac systolic function with a mean LVEF of 62%. Mean LVEF was, however, higher in the two other groups: 69 and 67%, respectively (p 0.0001). Left ventricular peak filling rate (LVPFR) and left atrial passive emptying volume (LApev) were significantly reduced in women with PPCM: mean LVPFR in the three groups was 229, 276 and 265 ml/s/m^2 (p=0.005) and mean LApev was 13, 19 and 20 ml/m^2 (p0.0001). Exercise capacity (peak VO_2) was also reduced in women with PPCM: mean peak VO_2 was 29.6, 43.2 and 45.4 ml/kg/min (p0.0001). Only one woman with PPCM had late gadolinium enhancement (LGE).

Conclusion

Women in this nationwide cohort generally recovered LV systolic function. Exercise capacity was however significantly reduced compared to matched control groups, which may be explained by impaired diastolic function in the PPCM group. Focal myocardial fibrosis assessed with LGE was uncommon in this cohort.

Recurrence of Congenital Heart Disease in Offspring's of Mothers with Congenital Heart Disease Screened by Fetal Echocardiography

Vlasta Fesslova[1], Annalisa Inversetti[2], Cristina Sigismondi, Cristina Sigismondi[2], Jelena Brankovic[1], Jelena Brankovic
[1]*Policlinico San Donato IRCCS, Italy*
[2]*Vita Salute Univiersity, Ospedale San Raffaele, Italy*

Background

Pregnant women with congenital heart disease (CHD) are at risk of recurrence. Preconceptional counselling is very delicate in this population and the data relative to the recurrence rates are of extreme use in this field.

Objective of the study

To analyze the recurrence of congenital heart disease (CHD) in offspring's of mothers affected with CHD.

Material and methods

Retrospective-prospective study of 310 pregnancies of 271 mothers with CHD studied by fetal echocardiography in our Center between Jan. 1995 and Dec.2015. Thirty nine women were followed-up during 2-3 pregnancies. Twenty seven women, all operated, had cyanotic CHD, in 34 pregnancies and 244 had acyanotic CHD, operated in 149, in 276 pregnancies. Thirty six had multiple familial risks (2-5 relatives)

Results

Twenty three probands had CHD (total recurrence rate 23/310 =7.4% pregnancies), 1/34 pregnancies with cyanotic CHD (2.9%), 22/244 pregnancies with acyanotic CHD (9%). When mother alone was affected, the recurrence was 21/235=8.9%; when mother and another relative were affected the recurrence rate was 2/36=5.5%. The specific recurrence was higher in VSD (7/71=9.8%), ASD II (considered at age 1yr, diam.8mm- 7/88=7.9%), aortic stenosis (3/34=8.8%), ductus arteriosus (1/12=7.7%) and in AVSD (2/4cases). Concordant lesions occurred in 9 cases, partially concordant in 7, discordant in 6 cases.

Conclusions

Our data confirm a relevant recurrence of CHD in affected mothers, despite the numerical limits of our population. This fact has to be taken in account in prenatal counselling.

Sustaining a Pregnancy Despite the Odds – High Risk LVNC Delivery

Bashaer Gheyath[5], David Majdalany[1], Katherine Singh[2], Peter Aziz[4], Kenneth Zahka[4], Edward Soltesz[3], Shinya Unai[3], Michael Tong[3], Eileen Hsich[1]
[1]*Cleveland Clinic Department of Cardiovascular Medicine, USA*
[2]*Cleveland Clinic Department of Obstetrics and Gynecology, USA*
[3]*Cleveland Clinic Department of Thoracic and Cardiovascular Surgery, USA*
[4]*Cleveland Clinic Department of Pediatric Cardiology, USA*
[5]*Cleveland Clinic Department of Internal Medicine, USA*

Background

Left Ventricular Noncompaction Cardiomyopathy (LVNC) is characterized by extensive trabeculations of the left ventricle. Clinically it is associated with embolic events, heart failure, and arrhythmias. This condition was previously viewed as a contraindication to pregnancy. There have been a few cases of pregnancies carried to term published in the literature. We report a successful pregnancy of a symptomatic patient with LVNC, acquired QT prolongation, and a history of ventricular fibrillation (VF) with subsequent implantable cardiac device (ICD) placement, who decided to maintain her pregnancy despite expert advice.

Case

A 21-year-old female with a history of LVNC diagnosed in childhood presented with VF in the setting of a drop in her LV ejection fraction to 32%. She was noted to have acquired QT prolongation with subsequent ICD implantation. A year later, she had an unplanned pregnancy and decided against termination. She was admitted at 21-week gestation for symptomatic heart failure which was complicated by ventricular fibrillation requiring external defibrillation and subsequent ICU admission with intra-aortic balloon pump support. Due to maternal instability, emergent C-section with extracorporeal membrane oxygenation (ECMO) backup was performed at 26-week gestation. Subsequently, she required a left ventricular assist device (LVAD) which was complicated by a large thrombotic stroke with successful thrombectomy. After a five-month admission, she was discharged to rehabilitation with improvement in her motor and neurologic function. Her newborn was in the NICU for complications related to prematurity with a patent ductus arteriosus requiring closure. Ultimately, the mother and child were discharged home well.

Conclusion

This case describes the complicated yet feasible course of LVNC in the setting of acquired LQTS and decompensation prior to 25-week gestational age. In addition to the importance of a planned multidisciplinary team approach, our case highlights the morbidities associated with a successful pregnancy in patients with a high-risk profile.

Successful Pregnancy and Delivery Following Two Heart Transplantations

Karen Florio[2,4], Emily Williams[4], Tara Daming[2,4], Anna Grodzinsky[1,3], Krishna Patel[3], Stephanie Sparkes[5], Laura Schmidt[1,3], Valerie Rader[1,3], John Lee[1,3], Rebecca Gray[2], Darcy White[2], John Spertus[1,3], Anthony Magalski[1,3]

[1]*Saint Lukes Hospital of Kansas City, USA*
[2]*Saint Lukes Hospital of Kansas City, USA*
[3]*University of Missouri-Kansas City, USA*
[4]*University of Missouri-Kansas City, USA*
[5]*Saba University, Netherlands*

Background

Pregnancy after heart transplantation is a challenging clinical scenario due to risks of rejection, embryopathy and cardiac changes in pregnancy.

Objective

We present a successful second pregnancy in a patient with a hereditary dilated cardiomyopathy and a history of two heart transplants.

Results

This is a case of a 37 year old G2P1001 with a pregnancy complicated by a history of two heart transplantations (1997 for presumed viral dilated cardiomyopathy and 2006 following cardiac allograft vasculopathy), tacrolimus-induced DM and hypertension. She was followed by her transplant team, congenital cardiology, endocrinology, and maternal-fetal medicine. A maternal echocardiogram was performed in the 1st trimester with normal ventricular size and function, no valvular abnormalities and an EF of 65%. Maternal echocardiograms were repeated serially throughout pregnancy without significant changes. She was continued on prednisone and tacrolimus, and her diabetes was well controlled. Detailed anatomic assessment and a fetal echo were performed and were normal. Throughout her gestation, the patient remained as NYHA class I. Serial fetal growth ultrasounds were normal and antenatal testing was initiated at 32 weeks. At 36 weeks, she was admitted for rupture of the membranes and repeat cesarean was performed without complications under regional anesthesia. She delivered a female infant, with APGARS of 8 and 9, weighing 2515g, with no neonatal complications. Her postpartum course was uncomplicated. Postpartum, she agreed to genetic testing for hereditary cardiomyopathies, which showed two mutations: LMNA, a known pathogenic variant for dilated cardiomyopathy, as well as DSP, a variant of unknown significance. Her offspring tested positive for the LMNA variant.

Conclusion

This is the first reported successful pregnancy in a patient who had undergone two heart transplants. While challenging, successful pregnancy outcomes following heart transplantation can be achieved by employing a multidisciplinary approach.

Pulmonary Hypertension in Pregnancy: The Anesthesiology Cohort

Marie-Louise Meng[1], Ruth Landau[1], Olof Viktorsdottir[3], Jennifer Banayan[4], Tamila Grant[4], Brian Bateman[2], Richard Smiley[1], Elena Reitman[1]
[1]*Columbia University Medical Center, USA*
[2]*Brigham and Woman's Hospital, USA*
[3]*Massachusetts General Hospital, USA*
[4]*University of Chicago, USA*

Background

Pulmonary hypertension (PH) carries a risk of maternal mortality. Anesthesiologists care for these patients during termination or delivery. We report our continued registry of patients with PH in pregnancy with the hope of recruiting other medical centers to contribute cases to this rare disease cohort.

Objective

The goal is to determine whether pregnancy outcomes differ depending on etiology and PH severity PH and to determine what treatments and delivery methods may improve outcomes.

Methods

The following information will be collected per case: WHO PH classification, NYHA classification, pulmonary vascular pressure, medical treatments, mode of delivery, anesthetic management, ECMO use, maternal mortality and fetal outcomes. Patient data will be de-identified and stored in REDCAP.

Results

Fifty-one women are in the cohort, 49 of whom were in our published report this year.(1) Mortality rate is 16%. The majority of deaths occurred in women with WHO group 1 PH. Seven women required ECMO support. There were 19 vaginal deliveries, 16 cesarean deliveries, 6 intrapartum cesarean deliveries, 7 terminations of pregnancy, one IUFD followed by a dilation and evacuation procedure and the delivery method for 2 women was unknown. Mortality rate was not higher among women that delivered vaginally but there were 2 deaths among the women who required intrapartum cesarean deliveries. Neuraxial anesthesia was performed in the majority of cases with no anesthesia-related adverse events. Preterm delivery was more common in women with severe compared with mild PH.

Conclusion

Adding women to this cohort will help clarify how maternal outcomes differ based on etiology and severity of PH. A large cohort is needed to allow evidenced-based conclusions on whether there is an optimal strategy for delivery in these women.

1. Meng ML, et al. Pulmonary Hypertension in Pregnancy: A Report of 49 Cases at Four Tertiary North American Sites. Obstet Gynecol. 2017;129(3):511-20.

Maternal Complications and Pregnancy Outcome in Women with Pulmonary hypretension: A Single Center Experience from South India.

Anish Keepanasseril[1], Ajith Ananthakrishna Pillai[2], Yavana Suriya J[1], Aswini R[1], Santhosh Satheesh[2], Pankaj Kundra[3], S Soundara Raghavan[1]

[1]*Jawaharlal Institute of Postgraduate Medical Education & Research, India*

[2]*Jawaharlal Institute of Postgraduate Medical Education & Research, India*

[3]*Jawaharlal Institute of Postgraduate Medical Education & Research, India*

Background

Pulmonary hypertension when associated with pregnancy carries a poor prognosis. Morbidity and mortality is still high in low to middle income countries in such patient where resources are limited and specialist services are segregated to few tertiary centers. Even though considered to be a contraindication to pregnancy, women with pulmonary hypertension presents late in pregnancy in these centers.

Objective

To study the pregnancy outcome and maternal complications in women with pulmonary arterial hypertension attending the tertiary center in south India from Jan 2011 to Jun 2017

Methods

This is an observational study analyzing the pregnancy outcome and maternal complications in pregnancy complicated with pulmonary hypertension. All patients with right ventricular systolic pressure (RVSP) 40mmHg at rest, measured by echocardiography or right heart catheterization, were included. Patients with elevated RVSP caused by outflow tract obstruction/pulmonary stenosis or secondary to left heart structural valvular disease were excluded.

Results

There were 4 maternal deaths (out of 50 patients) in the study period: three of them has idiopathic pulmonary hypertension and one had Eisenmengers syndrome. Mean at time of admission was 23.44 years and the mean gestation age at delivery was 37.6 weeks. Fifteen women (30%) received sildenafil during pregnancy. Mean RVSP was 68.11mm Hg (range 40-150). All had normal biventricular function with 2 of the m having right ventricular contractile dysfunction. Mean birth weight was 2.45 Kg and there were 11 perinatal death in the study.

Conclusion

With a multidisciplinary team approach, pregnancy outcome can be optimized in pregnant women with pulmonary hypertension. Women with iPAH has a higher risk of maternal and fetal mortality, so pre-pregnancy counselling and early booking in pregnancy should be encouraged even in developing nation to optimize the outcome.

The Predictive Value of N-Terminal pro-B-Type Natriuretic Peptide in Early Pregnancy For Cardiovascular Complications in Pregnant Women with Congenital Heart Disease

Anne Siegmund[1], Marlies Kampman[1], Barbara Mulder[2], Martijn Oudijk[3], Arie van Dijk[4], Krystyna Sollie-Szarynska[5], Gertjan Sieswerda[6], Steven Koenen[7], Caterina Bilardo[5], Dirk Jan van Veldhuisen[1], Petronella Pieper[1]

[1]*University Medical Center Groningen, University of Groningen, Netherlands*
[2]*Academic Medical Center, University of Amsterdam, Amsterdam, Netherlands*
[3]*Academic Medical Center, University of Amsterdam, Netherlands*
[4]*Radboud University Medical Center, Radboud University, Netherlands*
[5]*University Medical Center Groningen, University of Groningen, Netherlands*
[6]*University Medical Center Utrecht, University of Utrecht, Netherlands*
[7]*University Medical Center Utrecht, University of Utrecht, Netherlands*

Background
It is challenging to identify individual women with congenital heart disease (CHD) who will experience cardiovascular complications during pregnancy. N-terminal pro-B-type natriuretic peptide (NT-proBNP) levels at 20 weeks of pregnancy are of additional value to identify women who will suffer maternal cardiac complications later in their pregnancy. The predictive value of NT-proBNP in early pregnancy is unknown.

Objective
We aimed to investigate the predictive role of NT-proBNP levels in early pregnancy to predict cardiovascular complications later in pregnancy in women with CHD.

Methods & Results
We included 105 pregnant women with CHD from the prospective ZAHARA III study (Zwangerschap bij Aangeboren HARtAfwijkingen, pregnancy in congenital heart disease). Follow-up with clinical evaluation, echocardiography and NT-proBNP measurements were performed in early pregnancy (12 [2-17] weeks). Cardiovascular complications occurred in 4.8% of the women and were associated with elevated NT-proBNP levels in early pregnancy (β=4.09, p=0.002),the presence of a mechanical valve (β=2.77, p=0.008) and WHO3 class preconception (β=3.05, p=0.008). Figure 1 shows the distribution of patients with NT-proBNP levels and its relation to cardiovascular complications. The negative predictive value of NT-proBNP 235pg/mL was 98.9% (1 patient with abdominal aortic thrombosis, NT-proBNP was 50pg/mL) and the positive predictive value was 66.7%. Right ventricular function (tricuspid annular plane systolic excursion) preconception was associated with higher NT-proBNP levels in early pregnancy in a multivariable regression model (β=-0.041, p=0.045).

Conclusion
An increased NT-proBNP level in early pregnancy is indicative for cardiovascular complications later in pregnancy. For patients with low NT-proBNP in early pregnancy, less intensive cardiac follow-up during pregnancy may be considered. When NT-proBNP levels are elevated in early pregnancy, echocardiography with focus on right ventricular function is advisable to identify high risk pregnancies.

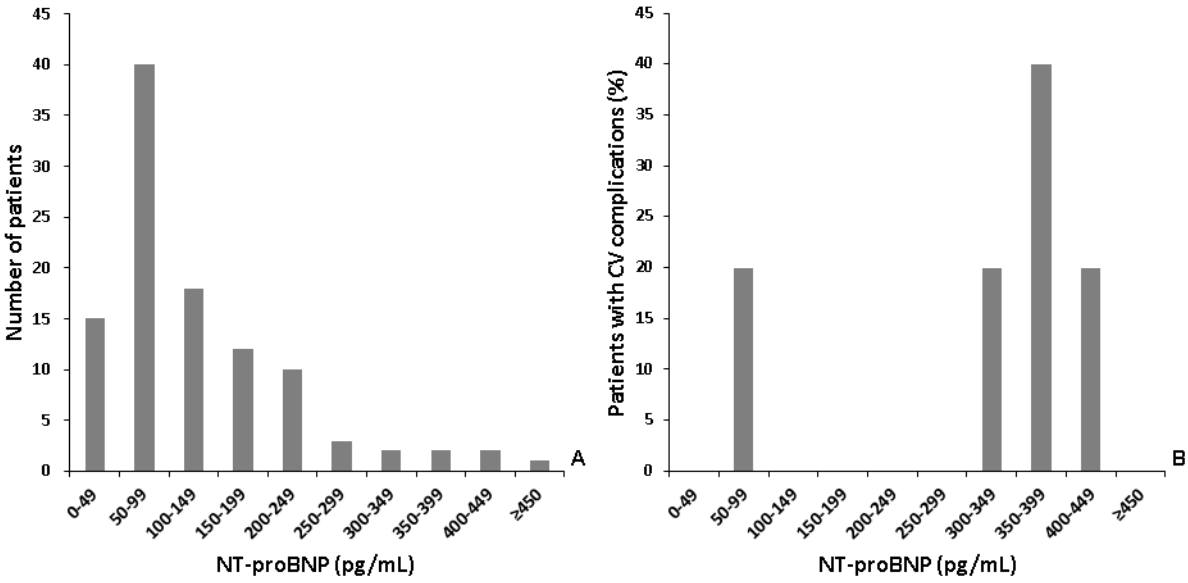


Figure 1. The distribution of patients by N-terminal pro-B-type natriuretic peptide level in early pregnancy (A), percentage of cardiovascular complications by N-terminal pro-B-type natriuretic peptide level (B). CV complications, cardiovascular complications; NT-proBNP, N-terminal pro-B-type natriuretic peptide.

Criss-Cross Heart in Pregnancy: A Case Report

Elisa Gherbesi[1], Cecilia Gobbi[1], Marco Schiavone[1], Mattia Squillace[1], Giovanni Malanchini[1], Margherita Calcagnino[1], Manuela Wally Ossola[2], Anna Maria Colli[1], Carla Bonanomi[1], Federico Lombardi[1]

[1]*Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, University of Milan, Italy*

[2]*Fondazione IRCCS Ca' Granda Ospedale Maggiore Policlinico, University of Milan, Italy*

Background

Criss-cross heart is extremely rare among congenital heart diseases (0.1%). It is characterized by an abnormal rotation of the heart on its long axis and often associated with other multiple defects. Optimum management and pregnancy outcome in women with criss-cross heart has not been described so far.

Case Presentation

We describe the case of a 33-years-old primigravida woman, with criss-cross heart and atrio-ventricular and ventricular-arterial concordance.

At the age of 4, she underwent complex surgery (atrial septal defect closure, arterial duct ligature and pulmonary artery enlargement with pericardial patch).

Her last evaluation before pregnancy (echocardiogram and cardiac magnetic resonance) documented normal biventricular function and absence of ventricular outflow tract obstructions.

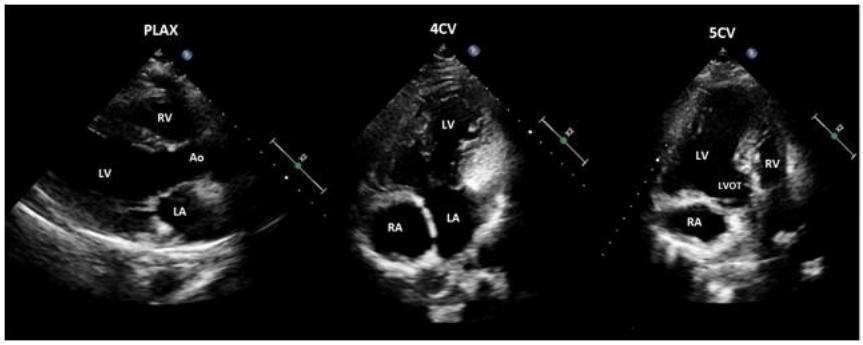
At presentation to our tertiary maternity centre, at 11 weeks of pregnancy, she was asymptomatic. An echocardiogram confirmed previous findings (Figure).

Thus, after a multidisciplinary evaluation that included adult congenital heart disease cardiologists and obstetricians, we settled a strict follow-up with monthly evaluations including echocardiogram, 24-hour ambulatory ECG and obstetric ultrasonography in order to detect as early as possible any complication (heart failure, tachyarrhythmia, atrio-ventricular blocks, and intrauterine growth retardation).

The patient remained asymptomatic until week 29th, when she was admitted to our hospital for threatened premature labor: she was treated with tocolytic and steroids for foetal lung development. At this time she developed symptoms and signs of heart failure (confirmed by elevated NT-pro-BNP values) and was successfully treated with intravenous furosemide.

At 37 weeks, for maternal request, an elective caesarean section was performed in spinal anesthesia, without complications.

A healthy baby girl of 3100 gr was delivered. A transient mild respiratory distress syndrome was quickly resolved with continuous positive airway pressure.



Conclusion

This case emphasizes the importance of a multidisciplinary team of obstetricians, cardiologists and neonatologists with expertise in high-risk pregnancies for a good maternal and foetal outcome.

Maternal Mortality in the Post-delivery Period in Patients with Pulmonary Hypertension

Vifan Lu, Cynthia Taub
Montefiore Medical Center, USA

Background

Pregnancy in women with pulmonary hypertension (PH) has long been regarded as high risk for maternal complications including death. This study is to investigate maternal mortality in the post-delivery period in patients with pulmonary hypertension and its risk factors.

Methods

Hospital discharge records from Statewide Planning and Research Cooperative System (SPARCS) in New York state between 2008-2014 linked to New York State death certificate were analyzed retrospectively. The study cohort consisted of PH patients with hospital records of labor and delivery. PH was identified with ICD-9 codes 415.0, 416.0, 416.8, 416.9, and further categorized into PH associated with congenital heart disease (CHD), PH associated with cardiomyopathy (CM) or valvular heart disease (VHD), and isolated PH (ICD-9 codes refer to Table 1). Labor and delivery was identified with ICD-9 codes 72-75, V27 or 650–659. Last delivery dates were retrieved with procedure date associated with delivery-related procedures.

Results

Among 1,098,694 patients with records of delivery, 656 PH patients were identified, including 68 PH patients with CHD, 358 PH patients with isolated PH and 230 PH patients with CM or VHD (Table 2). 20 deaths were found after delivery. The average age at death was 31.7 ± 7.5 years. 7 patients had mortality within 1 month of delivery, 10 within 6 months of delivery, and 10 deaths beyond 1 year. PH with CHD showed higher mortality within the first 6 months of delivery, while PH with CM or VHD showed higher mortality in the long term (Figure 1).

Conclusion

Mortality within 6 months of delivery in patients with pulmonary hypertension was not negligible and indicates close follow-up for a prolonged period is needed for the high-risk patient population. Subtypes of PH could be associated with different mortality patterns post-delivery.

Pregnancy in Patients with Mechanical Prosthetic Valves: Management Issues at a Tertiary Care Institution in North India.

Amrit Gupta[1], Varuna varma[2], Nirmal Gupta[2,3], Naresh kumar Bamil[2,4]
[1]*Sanjay Gandhi Post Graduate Institute of Medical Sciences, India*
[2]*Sanjay Gandhi Post Graduate Institute of Medical Sciences, India*
[3]*Sanjay Gandhi Post Graduate Institute of Medical Sciences, India*
[4]*Sanjay Gandhi Post Graduate Institute of Medical Sciences, India*

Pregnancy presents a unique challenge in women with prosthetic heart valves. Mechanical heart valves are inherently prone for increased thromboembolic events and acute hemodynamic problems due to mechanical dysfunction caused by valve thrombosis and pannus formations. Advanced heart disease associated with poor knowledge about pregnancy related issues, appropriate maternal care during pregnancy, malnutrition, also pose a greater challenge in these patients.

Between April 2002- till Mar 2017, 716 married young women aged between 19-35 years (child bearing age) received mechanical prosthetic valves at a single unit. Thirty four women died within 30 days following operation due to cardiac causes. In the remaining six hundred and fifty-six (682) women another 76 died or were lost to follow-up.

Of the remaining 606 survived beyond 1-year to be able to conceive a pregnancy. The median follow-up of 606 survivors (who are in regular follow-up) was 7.5 years. 112 out of 606 became pregnant during the period from 2006 to 2017 on 1 or multiple occasions. All women received only oral Nicoumalone as anticoagulant as standard of care. Sixty seven women had successful deliveries and rest 45 had single or multiple abortions. All women with the high risk situation were managed under supervision of obstetrician trained to manage high-risk pregnancies at a medical university hospital or at our MRH unit. nine children out of sixty seven had mild anticoagulation related embryopathy.

The overall incidence of complications in the most rigorously and despite focused management of young Indian pregnant patients with mechanical prosthetic valves is higher than compared to western standards but best achievable at tertiary level.

Our innovative management protocol, maternal anticoagulation regimen, maternal and fetal complications and outcomes associated in above mentioned scenarios in women with mechanical prosthetic valves will be discussed and presented.

Maternal and Neonatal Outcomes Following a Fontan Procedure: A 10-year Retrospective Study

Yalda Afshar[1], Mary Canobbio[2], Elizabeth Hoang[1], Jamil Aboulhosn[2], Brian Koos[1]
[1]*University of California, Los Angeles, USA*
[2]*University of California, Los Angeles, USA*

Background

Women with single ventricle physiology and Fontan procedure are surviving into adulthood. They are often discouraged from pregnancy, despite the paucity of data regarding outcomes.

Objective

We aimed to determine a relationship between maternal cardiac function and a small for gestation age (SGA) neonate.

Methods

This is a single-center, retrospective cohort study of women with a single ventricle physiology and Fontan procedure, who delivered over a ten-year period (2006-2016) at UCLA. All pregnancies exceeding 24-weeks' gestation were evaluated for differences in maternal and neonatal characteristics.

Results

Twenty-three women involving 26 pregnancies, were identified. Mean age at pregnancy was 25.1±4.1 years and mean BMI was 26.4±2.8 kg/m^2. The women were racially and ethnically diverse. There were no maternal deaths. The most common cardiac complications during pregnancy were sustained arrhythmia (36%) and decompensated heart failure (21%). Ejection fraction (EF) declined from pre-pregnancy to the first-trimester (p0.05). EF returned to pre-pregnancy levels within 1-5 years postpartum. Forty-six percent had a vaginal delivery, of which 58% were operatively assisted. Ninety-five percent had regional anesthesia. Fifty percent of neonates were born preterm (37-weeks). There were two intrauterine fetal demises (7.7%). Mean gestation age was 34 5/7±4 5/7 weeks with an average birth weight of 2291±545 grams and a 29% NICU admission rate. Most (77%) of the neonates were SGA at delivery (10thcentile). The reduction of first trimester maternal EF correlated with a SGA neonate (Spearman-rank correlation (r=0.604, p0.05)) adjusted for gestational age.

Conclusion

Women with a Fontan procedure have a significant decline in EF during the first trimester. The declining antenatal EF is predictive of a SGA neonate. Further research is needed to bridge the gap between components of cardiac reserve that can help us understand what modulates the underlying neonatal outcome.

The Late Effects of Pregnancy on Ascending Aortic Dimensions in Patients with Marfan Syndrome

Rafael Kuperstein, Amihay Shinfeld, Michal Simchen
Sheba Medical Center, Tel Aviv University, Israel

Background and aim

While the immediate effects of pregnancy in patients with Marfan syndrome (MS) have been evaluated, the late effects of subsequent pregnancies in these patients are less known.

For this purpose we retrospectively evaluated the changes in ascending aortic dimensions over time in women with MS who underwent pregnancy compared with women with MS who did not experience pregnancy.

Methods

Two groups of women with MS who were under care in our institution were compared. Group A included 19 women with MS who experienced 44 pregnancies. Group B included 19 patients with MS who never got pregnant. Eight women who underwent surgery before their first visit to our hospital and one patient, who developed post-partum type A dissection and underwent a Bentall procedure were excluded from the study.

Results

When compared with the patients in group B, patients in group A were older (37.8±4.9 vs.33±7 years, p=0.02) with longer follow-up time (6.4±2.8 vs 3.8±2.9 years). The mean number of pregnancies per woman was 2.3 (range 1-6 pregnancies). Baseline aortic diameters as well as the aortic diameters at the end of the follow up period were similar between groups (37±5 vs 33±9 mm and 38±5 vs 35±5 mm respectively, p-ns for both). The aortic diameter increased by 1.7±3 mm in group A and n 1.9±3 mm in group B (p=0.9). The percentual change in diameter was similar in both groups (5±8% in group A vs. 5±7% in group B, p=0.9) .Chronic medical treatment was similar in both groups (11 patients in each group).

Conclusions

Patients with MS who underwent pregnancies were older and had a longer follow-up time. Subsequent pregnancies were not associated with an increase in the rate of aortic dilation in patients with MS.

Nitrous Oxide: An Analgesic Option for Laboring Patients with Congenital Heart Defects

Peter Yeh[1], Anushree Agrawal[2], Hayley Schultz[4], Ahmed Kheiwa[2], Molly Killion[5], Nisha Parikh[2], Ian Harris[2], Juan Gonzalez[3], Jennifer Lucero[1]

[1]*University of California San Francisco, USA*
[2]*University of California San Francisco, USA*
[3]*University of California San Francisco, USA*
[4]*University of California San Francisco School of Medicine, USA*
[5]*University of California San Francisco, USA*

Background

Maternal cardiac diseases, including both acquired and congenital heart defects (CHD) are becoming more prevalent in obstetrics. Labor analgesia is often sited as one of the major concern of patients during labor. The epidural is considered the gold standard for labor analgesia, but nitrous oxide (mixture of 50-50 oxygen & nitrous) is an additional analgesic option. Many patients utilize nitrous as a bridge between early and active labor prior to having neuraxial analgesia. In general, nitrous oxide for labor analgesia is uncommon in the United States, with our institution being one of the few places to widely offer it as an analgesic option. There have been relatively few studies looking at the efficacy of nitrous oxide as a form of analgesia during labor for patients with CHD.

Objective

To evaluate nitrous oxide utilization and safety as an analgesic option in women with CHD during labor.

Methods

This was a descriptive study of patients with congenital and acquired cardiac disease who received care during their labor and delivery at a single institution from 2008-2017. History and modes of labor analgesia were abstracted from medical records.

Results

Of 340 pregnancies in women with cardiac disease, 150 were in women with CHD and 190 in those with acquired cardiac disease. Of all pregnancies, nitrous oxide was utilized in 41 (12%) with 20 (5.8%) pregnancies receiving both forms of analgesia (nitrous & epidural). In pregnancies among women with CHD, 126 (84%) received an epidural, 18 (12%) utilized nitrous and 9 (6%) used both nitrous prior to receiving an epidural. There were no significant morbidity or mortality events associated with nitrous oxide usage in either group.

Conclusion

It is possible to safely offer patients with CHD nitrous oxide as an option for labor analgesia.

Impella Assisted Cesarean Delivery in a 27 6/7 Weeks Gestation 30 Year Old Female with Newly Diagnosed Cardiomyopathy and Multifocal Incessant Ventricular Tachycardia.

Brendan Carry[1], Jason Costa[1], Alex Reyentovich[1], Shaline Rao[1], Stephen Pan[1], Mara Rosner[2], Jill Westcott[2], Francine Hughes[2], Catherine Weinberg[1]
[1]*NYU Langone Medical Center, USA*
[2]*NYU Langone Medical Center, USA*

Background

The role of mechanical circulatory support in advanced heart failure continues to evolve. We describe an unusual case of decompensated heart failure in a 30 year old woman at 26 1/7 weeks gestation who underwent percutaneous placement of an Impella CP to support successful cesarean delivery at 27 6/7 weeks gestation.

Case

A previously healthy 30 year old gravida 1 para 0, at 26 1/7 weeks gestation presented with progressive palpitations and worsening dyspnea with minimal exertion for several weeks. Although hemodynamically stable, telemetry revealed multifocal incessant ventricular tachycardia. She was placed on esmolol and lidocaine with a decrease in ectopy. Transthoracic echocardiogram revealed a dilated left ventricle with EF of 20% and severe mitral regurgitation. A heparin drip was started. She required furosemide for management of pulmonary edema and was transitioned from IV lidocaine to oral mexiletine. On day #3 Bromocriptine was initiated, however she became hypotension and it was discontinued. On day #12, a Swan-Ganz catheter showed PA pressure of 60/36 mmHg, PCWP of 35 mmHg and CI of 1.9-2.0 L/min/m^2 by FICK. Throughout her hospitalization her fetus was monitored. Given her low cardiac output, an Impella CP was placed percutaneously under general anesthesia and a cesarean delivery was performed on day #13 at 27 6/7 weeks. She remained on Impella support for 36 hours. Postpartum, she was restarted on bromocriptine and heart failure therapy. Cardiac MRI postpartum demonstrated severe biventricular dysfunction and nonspecific extensive patchy LGE of the LV. She was discharged on day #21.

Conclusion

Impella CP is a viable option during delivery for the gravid patient with decompensated heart failure.

Mode of Delivery in Pregnant Women with Underlying Heart Disease, a Retrospective Cohort Study

Matthew Cauldwell[1], Philip Steer[1], Adam Jakes[2], Suzanne Wallace[3], Gemma Malin[3], Monique Sterrenberg[4], Gemme Ulivi[5], Tom Everett[5], Mark Johnson[1]

[1]*Imperial College London, UK*
[2]*Guy's and St Thomas*
[3]*Nottingham University Hospital*
[4]*Southampton Hospital*
[5]*Leeds General Infirmary*

Background

Reports suggest that pregnant women with heart disease (PWHD) have higher rates of assisted delivery and caesarean section (both emergency and elective) although data from the European Registry of Pregnancy and Cardiac disease suggest that caesarean section without an obstetric indication confers no benefit for the mother or her baby. European Society of Cardiology guidance supports vaginal delivery for the majority of women with heart disease. To assess practice in the UK we collated data on mode of delivery from a large cohort of PWHD (both congenital and acquired) managed in six UK tertiary centres.

Methods

Data was collected from medical and obstetric notes on mode of delivery in pregnancies 24 weeks gestation. In four centres control data on women who delivered immediately before and immediately after each index pregnancy were available. First and subsequent births were analysed separately.

Results

Data was obtained on 1245 pregnancies in PWHD and 2422 controls. Baseline demographics including age, maternal height and weight and BMI were not significantly different. In first births in PWHD (n=653) cf controls (n=1348), 27.9% (182) cf 43.2% (582) had a spontaneous vaginal delivery(SVD) (Fishers exact p0.0001), 29.9% (195) cf 23.5% (317) had an assisted vaginal birth (p=0.0026), 24% cf 22.8% (307) an emergency caesarean section(EMCS) (p=0.611) and 18.2% (119) cf 10.5% (142) had an elective caesarean section (ELCS) (p0.0001). In second and subsequent births in women with heart disease (n=571) cf controls (n=1006), 44.8% (256) cf 67.1% (675) had a SVD (p0.0001), 13.3% (76) cf 11.4% (115) had an assisted vaginal birth (p=0.2965), 14.5% (83) cf 8.3% (83) had an EMCS (p= 0.0002) and 27.3% (156) cf 13.2% (133) had an ELCS (p 0.0001).

Conclusion

Pregnant women with heart disease have significantly lower rates of SVD and significantly higher rates of ELCS.

Outcome of Pregnant Women with Turner Syndrome

Anthonie Duijnhouwer, Roland van Kimmenade, Arie van Dijk, Janneke Timmermans
Radboud university medical center, Netherlands

Background

Pregnancy in women with Turner syndrome has been associated with aortic dissection, especially in the presence of congenital cardiac malformations and aortic dilatation. During pregnancy, aortic growth is presumed to be possible due to the influence of hormones and increased blood volume. Fast aortic growth is a risk factor for aortic dissection and therefore pregnancy in Turner women is classified as high-risk.

Objective

This study evaluates the ascending aortic growth and outcome during pregnancy in women with Turner syndrome.

Methods

Retrospective analysis of all pregnant Turner women visiting the outpatient clinic of the department of cardiology. Ascending aortic diameters were measured during the pre-pregnancy cardiac-MR and echocardiography. These were compared to echocardiography during pregnancy at 12, 20 and 32 weeks. Presence of cardiac malformation were registered.

Results

In total of 28 pregnancies in 27 women were evaluated. In 19 women, at least 1 pre-imaging before and during pregnancy was available. The mean age during pregnancy was 31.1±4.4 years. Oocyte donation was performed in 76% and spontaneous pregnancy occurred in 24%. No life-threatening complications occurred during pregnancy or shortly after delivery. The mean ascending aortic diameter at pre-pregnancy cardiac-MR was 26±3.6mm and on echocardiography was 26.5±3.6mm, Bland-Altman analysis showed a good agreement (no proportional bias). The mean ascending aortic diameter increase was 0.67±2.63mm (range 7mm). Although the low number of patients, the presence of bicuspid aortic valve (35% of the women) and oocyte donation showed no tendency for faster aortic diameter increase during pregnancy.

Conclusion

In this small study, women with Turner syndrome seem to have a low risk of life-threatening complications when at pre-pregnancy imaging a normal ascending aortic diameter is present. The aortic growth rate is within the variability of the echocardiographic measurement.

Pregnancy in Marfan syndrome: Genotype and Phenotype in Relation to Maternal and Fetal Outcomes

Linnea Baudhuin, Katrina Kotzer, Michelle Kluge, Erin Conboy
Mayo Clinic, USA

Background

Pregnant patients with Marfan syndrome (MFS) have been shown to have an increased risk for cardiovascular complications. Increased risk for aortic events has been associated with haploinsufficient *FBN1* variants in the general MFS population. A correlation between genotype and pregnancy outcomes has not been reported previously.

Objective

We investigated the risk for aortic dissection and pregnancy outcomes in 33 *FBN1* genotype-positive patients with MFS.

Method

The Mayo Clinic Electronic Health Record was searched to identify female patients whom had undergone genetic testing for MFS and whom were pregnant or had given birth. Additional patients were identified through a database search from a Mayo Clinic genetic testing reference laboratory.

Results

Of the 33 patients with a causative *FBN1* variant, 21 had a dominant negative (DN) variant and 12 had a haploinsufficiency (HI) variant. There were a total of 71 live births (46 DN and 25 HI) and 7 miscarriages (3 DN and 4 HI). Only 7/21 DN (33%) and 6/12 HI (50%) patients knew they had MFS at the time of pregnancy. A majority of the women (26/33; 79%) were not on losartan or beta-blocker therapy during pregnancy. The DN group had a miscarriage rate of 6.1%, compared to the HI group rate of 13.8%. There was 1 case of aortic dissection occurring at 27 weeks, resulting in a C section of a 31 week old baby. Another case of aortic dissection occurred in the immediate postpartum period, and resulted in maternal death. Both of the women with aortic dissections had haploinsufficient *FBN1* variants, and neither were managed for MFS during their pregnancy.

Conclusion

Although this data is from a small cohort, it suggests that pregnant women with HI *FBN1* variants have a higher risk for maternal and fetal complications during pregnancy.

Dissecting the Risk of Pregnancy in Turner Syndrome

Nejc Pavsic, **Katja Prokselj**
Department of Cardiology, University Medical Center Ljubljana, Slovenia

Background

Turner syndrome (TS) is a common chromosomal disorder, clinically characterized by short stature and infertility. It is a multiorgan disorder, affecting primarily endocrine and cardiovascular system. Spontaneous pregnancies in TS patients are rare, but conception is possible with assisted reproductive technologies. However, due to increased risk for aortic dissection during pregnancy, detailed cardiological evaluation before conception is required. Aortic size index ASI2.5 cm/m^2 or 2.0 cm//m^2 with an additional risk factor defines significant aortic dilatation and is considered an absolute contraindication for pregnancy.

Methods and results

Since 2007, a total of 31 patients with TS (average age 28.9±8.8 years, BSA 1.54±0.14 m2) have been referred to adult congenital heart disease clinic of Department of Cardiology, University Medical Centre Ljubljana for cardiovascular assessment. Twelve patients had associated arterial hypertension, 1 patient had diabetes or coeliac disease and seven had hypothyroidism.

Echocardiography and magnetic resonance (MRA) or computed tomography angiography (CTA) was done in all patients. Aortic valve was bicuspid in 19% of patients, 16% had aortic stenosis and 19% aortic regurgitation. Major arterial vascular malformations were common, 1 patient had coarctation of aorta, 2 aberrant right subclavian artery and 2 aberrant left carotid arteries. The mean ASI was 1.94 cm/m^2 (1.37–2.7 cm/m^2). According to echocardiographic and MR/CT findings 8 patients (26 %) had an absolute contraindication for pregnancy and were advised against it. Only one patient with ASI 1.67 cm/m^2 and without additional cardiovascular malformations decided for pregnancy, her ante and postpartum course was uneventful.

Conclusion

Every patient with TS requires thorough cardiovascular assessment. In our group of patients cardiovascular malformations are common, with bicuspid aortic valve being the most prevalent. A quarter of our patients had an absolute contraindication for pregnancy. Only one patient without contraindications decided to proceed with the pregnancy, which was uneventful.

Managing Delivery in Women with Congenital Heart Disease: Results from the Cuban National Programme for Pregnancy and Heart Disease

Cristina Kinsella[1], Sara Thorne[2], Paul Clift[2], Hsu Chong[3], Katie Morris[3], Lucy Hudsmith[2], Sarah Bowater[2], Roman Vasallo Peraza[4], Jesus Ernesto Perez Torga[4], Pedro Antonio Roman Rubio[4]

[1]*University of Birmingham, UK*
[2]*Queen Elizabeth Hospital Birmingham, UK*
[3]*Birmingham Womens Hospital, UK*
[4]*Institute of Cardiology and Cardiovascular Surgery, Cuba*

Purpose

To provide an update on the delivery outcomes achieved in congenital heart disease (CHD) patients by the Cuban National Program for Pregnancy and Heart Disease.

Methods

This was a single-centre retrospective study on a prospectively collected database from 1st January 2000 to 15th May 2017. Baseline data: demographics (age, province of origin), diagnosis, co-morbidities and functional status. Primary outcomes: duration of pregnancy, pregnancy outcome, any maternal or fetal complications.

Results

Overall 467 pregnancies in 462 women reached the third trimester. Median age was 25 (21-29) years, with 71% of cases originating from Havana. Patients presented in NYHA class I (97%, n=453) or II (3%, n=14), 6% (n=27) of patients had arrhythmias, 1% (n=4) had significant co-morbidities, and 1% (n=4) had impaired left ventricular function. One pregnancy resulted in stillbirth, with the rest (n=466) resulting in live births (n=469) of median birth weight 3200 (2880-3420) grams. Median duration of gestation was 39 (38.6-40) weeks. Sixty-six percent (n=307) of patients had a vaginal delivery; caesarean section rates were 34% (n=161). Maternal cardiovascular complications (0.5%, n=2), and obstetric complications (14%, n=67) did not result in mortality. Being small-for-gestational-age was the commonest complication (10%, n=48) in neonates, with zero mortality. Three neonates (1%) inherited congenital cardiac defects. Post-delivery NYHA functional status was class I in 84% (n=393) patients, class II in 16% (n=73) of patients and class III in one patient (0.5%).

Conclusion

Despite considerable resource constraints, in middle-income countries management of CHD pregnancies according to existing guidelines can achieve excellent outcomes.