

Heart and Lung Disorders Complicating Pregnancy and the Puerperium: Pitfalls in Practice & Lessons Learned

James Nello Martin Jr.¹, James Martin Tucker^{1,2}

¹Department of Obstetrics & Gynecology, University of Mississippi Medical Center, Jackson, Mississippi, USA

²Winfred L. Wiser Hospital for Women & Infants, University of Mississippi Medical Center, Jackson, Mississippi, USA

Email: jnmartinjrmd@gmail.com

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Abstract

Objective: To review a case series of 12 women with unexpected heart and lung disorders that occurred during pregnancy and the puerperium, describing for teaching purposes the pitfalls in practice and the lessons learned from this experience. **Materials & Methods:** We reviewed case files of women with medical and/or hypertensive complications of pregnancy that were evaluated for medicolegal defense purposes by the first author between 1986-2015. Twelve women in these case files experienced unexpected cardiovascular and/or cardiopulmonary complications late in pregnancy or early in the puerperium. For each case, the pertinent medical record information was extracted. Important concepts as lessons learned are summarized and referenced for teaching purposes. **Results:** Five women had undetected preexisting heart disease which acutely deteriorated during the third trimester, four women developed postpartum heart failure related to pregnancy and delivery, and three women suffered an intrapartum cardiac arrest; none survived. Their case presentations illustrate the importance of obstetric health care professionals being alert to the signs/symptoms of developing cardiopulmonary disease late in pregnancy and following delivery so that timely evaluation and intervention can be accomplished to potentially avoid morbidity and mortality. Diagnostic categories include peripartum heart failure, high-risk chronic hypertension, superimposed preeclampsia, amniotic fluid embolism, pulmonary embolism and Raynaud's with occult pulmonary hypertension. **Conclusion:** These cases illustrate the diversity of ways that cardiovascular disease can suddenly complicate pregnancy and the early postpartum period. A major part of any effort

to enhance safer motherhood is a heightened awareness by obstetric specialists to consider the possibility of heart disease in all maternity patients so that appropriate consultation and collaboration with other specialists might help avoid major maternal morbidity or mortality.

Keywords

Cardiopulmonary, Cardiovascular, Maternal Morbidity & Mortality, Pregnancy & Postpartum Complications

1. Introduction

The 2018-2019 ACOG Presidential Task Force on Pregnancy and Heart Disease resulted in the 2019 publication of the ACOG Practice Bulletin on this important clinical issue [1]. Its impact upon enhanced pregnancy and postpartum care by obstetric specialists in the United States has yet to be measured, but its importance to obstetric practice is underscored by the sobering statistic that cardiovascular diseases are responsible for 26.5% of U.S. pregnancy-related deaths [2]. Cardiovascular disease affects approximately 1% - 4% of the nearly 4 million pregnancies in the United States each year, and it has increased significantly by 24.7% in recent years [3]. Acquired heart disease during pregnancy is increasing [4], contributing most to the recent rising trend in maternal cardiovascular disease-related deaths [5]. Key to reducing maternal mortality and major maternal morbidity in the United States is the need to improve patient outcomes for women who develop any form of new-onset heart disease during pregnancy and postpartum.

Diagnosis of maternal acquired heart disease during pregnancy and the postpartum period can be challenging because the overlap of cardiovascular symptoms with those of normal pregnancy can lead to delays in diagnosis and subsequent treatment. It is estimated that a quarter or more of maternal deaths could be prevented if CVD were routinely considered in the differential diagnosis by treating health care professionals [5] [6] [7]. Like maternal mortality and morbidity reviews conducted by states and the Centers for Disease Control (CDC), aggregated medicolegal reviews focused on specific issues can provide worthwhile information to assess potential pitfalls in practice and lessons to be learned to improve future practice. Although selection bias toward catastrophic results is inherent in this type of undertaking, it is nevertheless a powerful teaching tool. Thus our purpose in this investigation of 12 cases is to explore how cardiovascular/acquired heart disease can present to the obstetric health care professional, to determine how often one or more of the 4 key risk factors (Black, hypertensive, obese, maternal age > 40) for heart disease in pregnancy are present, and to make a determination if consideration of the diagnosis of CVD might have averted the outcome.

2. Materials and Methods

Among the 161 pregnancy-related medicolegal cases the first author reviewed during a span of 35 years, 12 involved women who experienced unexpected cardiovascular/cardiopulmonary complications during pregnancy and the puerperium. Clinical summaries and datasheets prepared by the first author from medical record analysis (all non-medical, legal-related materials had been shredded after case resolution) were reviewed critically by both authors. In addition, important learning/teaching points for consideration by obstetrician-gynecologists to enhance future patient care were summarized in bullet points for each case. Once the initial reviews and data sheets were completed for each study subject, the information was reviewed confidentially by the second author to assure accuracy of the recorded findings. Findings from this analysis are grouped according to time of event onset as antepartum, intrapartum or postpartum. Referencing of recent and relevant publications pertinent to each discussed diagnostic entity completes each case summary.

3. Results

Pregnancy and postpartum care for the 12 women in this report spanned the years 1988-2016. Maternal age ranged from 20 - 42 years (mean = 31.2 years); only 2 were older than 40. Seven of 12 were black, 4 were white, 1 was Asian. Hypertension either acute-onset or chronic was present in 8 of 9 antepartum or postpartum event women; none of the 3 intrapartum event patients were hypertensive. Regarding maternal obesity/weight, 10 of 12 exceeded 180 lbs in delivery weight (range 150 - 253 lbs, mean = 202.2 lbs). Gestational age at delivery ranged between 30 - 41 weeks (mean = 37.5). Four of the 9 antepartum/postpartum event patients were postpartum in time of onset. Every woman in this case series had one or more of the 4 high risk factors for cardiovascular/cardiopulmonary compromise in pregnancy; 5 of 12 had three of four high risk factors. Most women were nulliparous; three were parous. There were no survivors.

3.1. Antepartum

***Patient 1** developed shortness of breath at rest, pulse of 120/min, increasing edema, and widening pulse pressure with increasing systolic and decreasing diastolic pressures at 32 weeks' gestation not supportive of a preeclampsia diagnosis. Chest pain occurred with deep breaths, EKG did not indicate cardiac ischemia. Over the next month systolic blood pressures increased to a severe level, diastolic blood pressures decreased, pulmonary edema developed and heart rate exceeded 100/minute while cold medicines and antibiotics were prescribed without patient benefit. During multiple prenatal visits and emergency room visits, the patient was erroneously reassured that her symptoms were normal for pregnancy. Primary cesarean delivery under general anesthesia was augmented with multiple diuretic administrations of furosemide, the patient became normotensive only during surgery. She couldn't be weaned off oxygen after surgery*

and developed worsening respiratory distress, resuscitation after cardiac arrest on the third postoperative day was unsuccessful. In addition to extensive pulmonary edema and pleural effusions at autopsy, mild to moderate atherosclerosis and an enlarged left ventricle was discovered. Arrhythmia related to ischemic heart disease was assigned as cause of death.

Patient 1 Perspective: Pregnancy in women with pre-existing ischemic heart disease (IHD) is rising in frequency [8] [9] and renders the patient at risk for deterioration during pregnancy and postpartum. Complications are common especially in the presence of coronary atherosclerosis [10]. Risk for maternal mortality ranges between 2% - 8% [11] [12] and is rising in young women between the ages of 35 - 54 [13] [14]. There is almost a 50/50 split between non-ST-segment elevation and ST-segment elevation associated with myocardial infarction [8]. Increased cardiac work secondary to pregnancy can unmask underlying vascular disease. Lessons learned include:

- Every pregnant or postpartum patient with chest pain or cardiac symptoms should have urgent/emergent consideration of acute coronary syndrome (ACS), a condition which implies suspicion of myocardial oxygen deprivation culminating in myocardial injury and necrosis [15]. ACS can be caused by coronary atherosclerosis, dissection, embolism, spasm, arteritis and coronary artery occlusion related to aortic dissection. Expert cardiology supervision of heart care is important, preferably including echocardiography and consideration of other invasive and non-invasive testing such as troponins and B-type natriuretic peptide.
- Vital signs showing tachycardia with pulse ≥ 120 requires prompt evaluation to determine if underlying heart disease is present, as does the development of shortness of breath at rest [1]. Neither of these occurs in normal pregnancy; reassurance without adequate evaluation is ill-advised.
- Inability to wean a patient off oxygen following general anesthesia requires prompt evaluation for underlying cardiac and/or pulmonary disease.

Patient 2 had a strong family history of cardiovascular disease (CVD) and many years of labile chronic hypertension prior to achieving pregnancy in addition to complaints related to ear and neck pain unresponsive to large doses of acetaminophen. While taking antihypertensives to control her blood pressure during gestation, she developed asymmetric head-sparing fetal growth restriction in addition to recurring neck and chest pain which she did not report to health care professionals. At term the patient declined induction of labor. Several days later the undelivered patient died at home while sleeping. Autopsy revealed concentric left ventricular hypertrophy, arteriosclerosis, aortic tear and cardiac tamponade without evidence of aortic dissection.

Patient 2 Perspective: Chronic hypertension in pregnancy is increasingly common [16]. Additional benefit to thorough baseline heart and kidney evaluation early in pregnancy is the early identification of mothers with increased cardiac disease risk especially in those that have had poorly controlled hypertension

for years and disease-onset prior to age 30 [17]. Left ventricular diastolic dysfunction is frequently present in chronic hypertensives [18] especially if obese [19]; women with hemodynamic evaluation revealing increased systemic vascular resistance and low cardiac output are at high risk to develop superimposed preeclampsia [20] [21]. Stratification early in pregnancy of chronically hypertensive women based on the etiology of their hypertension, baseline work-up and comorbid medical conditions has been proposed as a better way to manage these pregnancies and reduce adverse outcomes [22]. Lessons learned include:

- Patients with long-standing chronic hypertension are at risk to have significant underlying heart pathology which should be evaluated antepartum by maternal echocardiography in addition to management by a Pregnancy Heart Team which includes a maternal-fetal medicine subspecialist [1].
- The development of acute chest pain in a pregnant or postpartum patient, as noted previously, should be quickly evaluated by a cardiologist using appropriate laboratory testing and imaging of the coronary vasculature.
- Consideration for delivery sometime early in the 37 - 39 week gestational age window, not electively deferring delivery past 39 weeks, is advisable especially in chronic hypertensives with longstanding disease and/or co-morbidities (“high risk”).

***Patient 3** had long-standing chronic hypertension and had been prescribed antihypertensive medication since adolescence. Her dosages were increased multiple times until 34 weeks’ gestation when evidence of superimposed preeclampsia led to hospitalization. Despite multiple unsuccessful efforts to control severe systolic hypertension and fetal monitor findings concerning for possible fetal compromise, the patient remained undelivered and was transferred out of labor and delivery to the antepartum floor. Early the next morning the patient was returned to the labor and delivery unit for cervical ripening and induction of labor at which time fetal heart motion was undetectable. Soon thereafter the patient became dizzy and suffered a cardiopulmonary arrest. Multiple attempts at intubation were unsuccessful while cardiopulmonary resuscitation proceeded and emergent cesarean delivery was initiated. During these efforts the patient aspirated gastric contents and had a complete placental abruption. Subsequently cardiac enzymes returned as very abnormal suggesting heart injury, acute kidney injury was also demonstrated but a dislodged endotracheal tube days later with airway loss led to death.*

Patient 3 Perspective: Preeclampsia superimposed upon chronic hypertension/vascular disease affects 25.9% [23] of pregnant chronic hypertensives, potentially aggravating the diastolic dysfunction, left ventricular stress and increased peripheral resistance already present [24]. Greater severity of preeclampsia raises the overall risk of severe cardiovascular morbidity during hospitalization for delivery [25]. Dramatic hemodynamic changes and fluid shifts characterize the immediate postpartum period in these patients [26]. Lessons learned are:

- A patient with chronic hypertension and superimposed preeclampsia is al-

ways a candidate for delivery when she reaches 34 weeks' gestation at which time expectant management or deferred delivery no longer has a place in management in the presence of complications such as fetal compromise and sustained severe hypertension unresponsive to therapy.

- The risk for placental abruption in a patient with chronic hypertension is enhanced in the presence of sustained severe systolic hypertension and in patients with longstanding, severe hypertensive disease involving the uterine vasculature.
- Patients with difficult to control severe blood pressure that require repeated systemically administered medications belong in labor and delivery or other intensive care settings, not the inpatient floor setting where more routine care is being administered.

***Patient 4** experienced considerable third trimester weight gain, edema, tachycardia in association with criteria to merit a diagnosis of preeclampsia at 35 weeks' gestation. However, admission was deferred and return evaluation was planned to occur in 2 weeks. One week later, the patient returned complaining of shortness of breath, palpitations, and tachycardia plus evidence of severe hypertension and 2 - 3+ proteinuria. Vaginal delivery was accomplished. Fluid intake exceeded urine output intrapartum and postpartum while tachycardia with shortness of breath persisted. Despite this, the patient without workup for probable heart disease was discharged home only to return the following morning at which time she suffered a cardiopulmonary arrest in the emergency room and could not be resuscitated. Autopsy was declined by the family; peripartum cardiomyopathy was suspected.*

***Patient 5** developed evidence of severe preterm preeclampsia (early-onset preeclampsia) with sustained severe hypertension and more than 20 pounds of weight gain over the preceding 4 weeks. Although proteinuria was present, liver enzymes and the patient's platelet count were normal. The patient, however, had a very low serum albumin. Approximately 48 hours after the initial betamethasone regimen was given for fetal indication, blood pressures returned to the severe range and antihypertensives were increased. The diagnosis was considered to be "mild PIH" and discharge home the following morning was contemplated. A severe headache that night was treated with morphine instead of magnesium sulfate. Early the next morning on the inpatient floor the patient was found unresponsive. Attempted resuscitation and emergent cesarean delivery were unsuccessful and the patient died. Autopsy revealed evidence of right heart failure with hepatic congestion, acute pulmonary and cerebral edema.*

***Patients 4 and 5 Perspectives:** Peripartum cardiomyopathy (PPCM) occurs every 3000 - 5000 pregnancies as cardiac failure with the left ventricular ejection fraction (LVEF) less than 45% in the absence of underlying heart disease or any identifying cause, developing in the last month of pregnancy or within 5 months of delivery [27] [28]. Although uncommon, the incidence of PPCM is rising [29]. PPCM and preeclampsia frequently occur together [30], possibly sharing*

pathophysiology and challenging diagnosis and management [31]; differentiation requires echocardiography examination of the mother [32]. Lessons learned include:

- Any patient with preeclampsia likely complicated with cardiopulmonary compromise by definition has severe disease requiring full evaluation by appropriate consultants and echocardiography to rule out the unlikely but possible occurrence of PPCM as a cause of maternal compromise and a risk to her health.
- A pregnant or postpartum patient with shortness of breath at rest requires prompt cardiac evaluation.
- Late gestation pregnant or postpartum women in the first months after delivery who present with shortness of breath, chest discomfort, palpitations, arrhythmias or fluid retention should be evaluated for peripartum cardiomyopathy
- Severe systolic acute-onset hypertension is an obstetric emergency requiring systemically antihypertensives given immediately to prevent stroke [33] [34] [35].
- Early-onset preeclampsia places a patient at greater risk for rapid disease progression and fetal compromise compared to late-onset preeclampsia; it is best managed as an inpatient due to the possibility of rapid progression [36].
- A severe headache in a patient with probable preeclampsia regardless of early-onset or late-onset type represents a risk for deterioration to eclamptic convulsion which is best managed with intravenous magnesium sulfate, close observation in labor and delivery, and consideration for delivery.

3.2. Intrapartum

Patients 6, 7 and 8 were at term in labor when a sudden deterioration in fetal status by monitor tracing occurred in association with uterine hypertonus, spontaneous amnion rupture, increased lower abdominal and chest pressure, and patient complaints of suddenly feeling hot, anxious, and short of breath. One patient was delivered emergently by cesarean delivery accompanied by excessive generalized bleeding without evidence of placental abruption. Immediately following surgery worsening hemorrhage and DIC led to heroic efforts to transfuse and embolize both hypogastric vessels by interventional radiology. Nevertheless shock led to severe encephalopathy and brain death followed by withdrawal of life support. In Patient 7 evidence of non-clotting peripheral blood samples soon after cardiopulmonary arrest with suspected DIC led to a decision to not operate and, despite aggressive resuscitation and blood product administration, the patient died. In Patient 8 emergent perimortem cesarean was delayed while resuscitation efforts were begun and cessation of maternal cardiac activity was awaited prior to initiating cesarean surgery. In two of three patients' autopsy evidence of amniotic fluid embolism (aggregates of occlusive squamous debris) in association with variable amounts of pulmonary edema, ascites, hemorrhage, cerebral edema and infarction, renal cortical/tubular hemorrhage, adrenal he-

morrhage and liver necrosis was found. In the third patient probable amniotic fluid embolism was considered to be the cause of death given the clinical circumstances of sudden shortness of breath in labor and cardiopulmonary arrest without evidence of any other etiology.

Patients 6, 7 and 8 Perspectives: Amniotic fluid embolism is an unpredictable, unpreventable obstetric emergency with an incidence of approximately one in 40,000 - 50,000 deliveries [37] [38] [39]. It is characterized by the sudden passage of amniotic fluid and fetoplacental cellular debris into the maternal circulation that incite a response and injury similar to the systemic inflammatory response syndrome (SIRS). Hypoxia, dyspnea, cyanosis, loss of consciousness and seizure-like activity with cardiac arrest accompany fetal heart rate abnormalities, uterine atony and bleeding. Maternal treatment is primarily supportive with prompt delivery in the undelivered woman with cardiopulmonary arrest; emergent trans-thoracic echocardiography is recommended to detect and guide therapy for a failing right ventricle [40] while coagulopathy is aggressively managed. Mortality ranges between 20% - 60% [37] [38] [39] [40] [41]. Lessons learned include:

- Perimortem cesarean delivery, also termed resuscitative hysterotomy [42], is imperative in the undelivered patient with suspected AFE since chest compressions during cardiopulmonary resuscitation in an undelivered term pregnant patient likely can produce only an estimated 10% of cardiac output [43].
- Emergent left lateral manual uterine displacement is advisable supplemented by tilting the bed or placing a wedge [44].
- Deferring initiation of resuscitative hysterotomy until a failing maternal heart arrests in the presence of a probable AFE is not advantageous either to the mother or fetus even if the 4 or 5 minute rule to accomplish delivery is possible [1].
- Simulation exercises to prepare for the emergent need for perimortem cesarean are ideal and strongly recommended for physicians who deliver obstetric care since AFE is infrequent and, when it happens, there is little time to respond appropriately—it is a strong reason to practice this rarely needed surgery in a safe simulation setting using the guidance of the ACOG PB on Heart Disease in Pregnancy section on perimortem cesarean/resuscitative hysterotomy [1].

3.3. Postpartum

Patient 9 was discharged home following a normal pregnancy course and cesarean delivery for dystocia. She was considered to have mild antepartum gestational hypertension the last week of gestation; orders for discharge were arranged early on the third postoperative morning after which two blood pressure readings above 150/90 were obtained but not reported to the physician before the patient left the hospital. Two days later while at home the patient experienced an episode of chest pain and shortness of breath with a self-measured

blood pressure reading showing severe systolic and diastolic hypertension. The following day she was given oral antihypertensive and diuretic agents to take at home but no further testing for cardiac troponins or otherwise was requested. Several days later the patient was found in asystole at home, cardiopulmonary resuscitation was unsuccessful. A myocardial infarction was found at autopsy associated with atherosclerosis, no evidence of dissection or pulmonary embolus was found.

Patient 9 Perspective: The immediate postpartum period is a high risk time for cardiovascular/cardiopulmonary compromise [1] [45]. The call for routine risk assessment for CVD, patient education and early, enhanced targeted postpartum follow-up as advocated in ACOG PB HDP is important to minimize maternal morbidity and mortality [1] [46]. Lessons learned include:

- The onset of hypertensive readings postpartum at or above 150/90 require further evaluation for underlying disease requiring further evaluation or the initiation of *appropriate* treatment using antihypertensives, blood pressure monitoring and re-evaluation by a health professional within 7 - 10 days of delivery [46].
- Sudden acute-onset sustained severe systolic and/or diastolic hypertension during *pregnancy* or postpartum, increasing patient risk for stroke and heart/vascular complications, is an obstetric emergency requiring immediate evaluation and management [47] [48] [49].
- The onset of chest pain especially with other findings such as shortness of breath requires immediate evaluation and management in a pregnant/*postpartum* patient especially if she has high risk factors for CVD (black race, hypertension, obesity or age over 40).

Patient 10 is a chronic hypertensive that received antihypertensive therapy throughout gestation while developing symmetrical fetal growth restriction. Late in gestation she developed superimposed preeclampsia and underwent cesarean delivery under general anesthesia. Six days postoperative she returned after discharge 3 days earlier, now with severe pulmonary edema and respiratory distress. She became pulseless while in transit by ambulance back to the hospital during which multiple unsuccessful attempts at intubation occurred before respiratory support was restored. Severe systolic and diastolic hypertension required emergency treatment even as a possible eclamptic convulsion occurred while the patient was being imaged. After magnesium sulfate and furosemide were ordered and given, echocardiography findings didn't support a diagnosis of peripartum cardiomyopathy. With intensive medical management for congestive heart failure as a complication of chronic hypertension plus superimposed preeclampsia, the patient recovered but evidence of anoxic encephalopathy persisted. The patient survived until sepsis, pneumonia and respiratory failure ended her life.

Patient 10 Perspective: The same hemodynamic constraints and compromise continue into the puerperium in women with longstanding chronic hypertension [24] [25] [26]. Individual patient-specific follow-up consistent with CVD

risk, co-morbidities and presence of high risk factors for heart disease should drive postpartum care as it should drive antepartum care [1] [22]. Lessons learned include:

- Any patient with longstanding chronic hypertension who becomes pregnant and requires **antihypertensive** therapy is at risk to have significant underlying heart disease; transfer to a higher level of care where delivery and postpartum can be optimally managed should be considered.
- Whenever there is a **possibility** of peripartum cardiomyopathy, echocardiography is indicated [29] [30] [31] [32].

***Patient 11** developed preeclampsia at term (late-onset preeclampsia) and underwent primary cesarean delivery. No prophylactic measures to lessen the risk of venous thromboembolism were utilized before or after surgery. After refusing transfusion recommended to treat her severe anemia, the patient was discharged home with reappearance 48 hours later complaining of new-onset bloody sputum, low grade temperature, acute onset shortness of breath and dyspnea on exertion. Severe diastolic hypertension was observed in association with pitting lower extremity edema. Believing this to represent unresolved preeclampsia complicating chronic hypertension, the clinician elected to use furosemide to off-load considerable fluid and infuse several units of packed red cells. Chest radiograph revealed mild cardiomegaly with evidence of residual vascular engorgement. The patient experienced recurring leg soreness, leukocytosis, sinus tachycardia; EKG revealed nonspecific T-wave abnormalities with possible lateral ischemia. There was no testing for troponins or evaluation for possible pulmonary embolus. Although leg cramps continued in both legs especially while walking and there was gluteal soreness, the patient was discharged home. Several days later at home she suddenly collapsed, vomited, became cyanotic and pulseless without respiration. Autopsy revealed the presence of a large established saddle pulmonary embolus.*

***Patient 11 Perspective:** One in 10 pregnancy-related deaths in the United States is due to pulmonary embolism [50]. Utilization of postpartum thromboembolism prophylaxis during delivery hospitalization varies among hospitals [51] [52] and frequently is overlooked even in patients at high risk due to recent cesarean surgery or having a preeclampsia-spectrum disorder. All strategies and consensus bundles rely on risk factor identification and thromboprophylaxis for the highest risk patients [53] [54] [55] [56] [57]. As a minimal approach the Society for Maternal-Fetal Medicine recommends sequential compression devices for all women who undergo cesarean deliveries with continuation until the patient is fully ambulatory [57]. A diagnostic and management strategy for pregnant and postpartum women is proposed based on clinical probability, D-dimer measurement, lower limb compression ultrasonography, and computed tomography pulmonary angiography [58]. Lessons learned are:*

- The possibility of pulmonary embolus should always be considered in any postoperative **patient**, especially one who was pregnant, had preeclampsia and had a cesarean delivery.

- Any postoperative cesarean delivery patient presenting with the acute onset of bloody sputum, **acute** shortness of breath and dyspnea on exertion should be considered to have, and evaluated for, a pulmonary embolus until proven otherwise.

***Patient 12** entered pregnancy with a known history of systemic lupus erythematosus (SLE) associated with Raynaud's disease. She had experienced two prior episodes of lupus pericarditis and multiple joint issues requiring hospitalization several years preceding this first pregnancy. No activation of SLE occurred during pregnancy nor were corticosteroids prescribed. After a failed cervical ripening attempt, the patient underwent cesarean delivery under general anesthesia without complication. A postoperative temperature spike was accompanied by rales bilaterally and night sweats which were considered to represent SLE or infection; antibiotics were initiated for presumptive endomyometritis while consultants pondered and tested for the possibility of SLE. By postoperative day 4 there was some mild persistent peripheral hypotension accompanied by shortness of breath, low urinary output and laboratory indices of acute kidney injury. After transfer to intensive care, echocardiography and EKG revealed evidence of right ventricular failure, severe pulmonary hypertension with tricuspid regurgitation; labs revealed hypoglycemia, metabolic acidosis and DIC to support a diagnosis of liver necrosis and failure. Lung scan was suspicious for pulmonary emboli. Despite heparin, high dose corticosteroids and hemodialysis, the patient deteriorated and died the following day. Autopsy was declined; longstanding occult pulmonary hypertension was suspected.*

***Patient 12 Perspective.** As recently as 2017, progress toward international consensus regarding diagnosis and management of Raynaud's phenomenon was not well advanced particularly regarding pregnancy management [59]. Raynaud's is a component of undifferentiated connective tissue disease at risk for systemic sclerosis which leads to mild biventricular systolic and diastolic dysfunction [60] [61] which in turn can be associated with vascular disease of the small pulmonary arteries that leads to pulmonary arterial hypertension (PAH). A leading predictor of PAH presence in patients with SLE is Raynaud's [62]. Pregnant SLE patients are at great risk of dying due to undiagnosed PAH. Thorough evaluation of a patient with SLE and Raynaud's for the presence of occult PAH is thus advisable prenatally and, if present, may form the basis of a recommendation to not undertake pregnancy due to its risk to decompensate into right ventricular failure [63]. Adenosine stress cardiovascular magnetic resonance (CMR) is advised to determine the myocardial perfusion reserve index (MPRI) and assess myocardial perfusion in the nonpregnant Raynaud's patient. [64] The lesson learned is:*

- The presence of PAH is a contraindication to pregnancy and, therefore, it is desirable to assess any SLE/systolic sclerosis/mixed connective disease disorder/Sjogren's patient prior to pregnancy. If pregnancy occurs, multidisciplinary diagnosis and management by a Pregnancy Heart Team as outlined in the ACOG PB on Pregnancy & Heart Team is advisable [1].

4. Discussion

The rising trend in maternal deaths related to cardiovascular disease appears to be due primarily to acquired heart disease, unknown and/or silent until late in pregnancy or in the days and weeks immediately following delivery. Many “acquired heart disease” maternal deaths are deemed to have been potentially preventable due to either clinician issues, patient issues, access issues or a combination of these. Clinician issues include the challenges to sustaining a high level of suspicion and expectation that any pregnant or postpartum patient can have a significant underlying cardiac disorder especially if they exhibit any of the four key heart disease risk factors of black race, obesity, hypertension or age over 40. Differentiating between a patient’s symptoms as normal or abnormal and thus urgently in need of cardiac evaluation is critical (see Table 2 in ACOG Practice Bulletin Heart Disease in Pregnancy) [1]. The other particularly valuable algorithm directing CVD assessment in pregnant and postpartum patients is Figure 1 in the 2019 Practice Bulletin; it uses patient symptoms, vital signs, risk factors and physical findings to initiate and direct the course of evaluation [1]. Testing (electrocardiogram) for potential heart pathology is warranted during pregnancy or postpartum in women that develop shortness of breath, chest pain or palpitations associated with maternal tachycardia. Normal or low levels of B-type natriuretic peptides (BNP) [65] are useful to exclude cardiac decompensation during pregnancy as are normal troponin levels which are specific and sensitive biomarkers of myocardial injury. D-dimer is not recommended as part of routine evaluation of cardiac disease in pregnancy or the postpartum period with the exception of assessing for pulmonary embolism [1] [58]. Computed tomography should be performed in pregnant or postpartum women presenting with chest pain when pulmonary embolism or acute aortic dissection is suspected. Echocardiography is particularly important to undertake in late gestation and postpartum patients developing nonischemic heart failure with a left ventricular ejection fraction of less than 45%.

In this era of safety bundles, there remains a place of major importance for obstetric specialists to be alert during pregnancy and postpartum care for any patient, particularly one with key risk factors to develop heart disease, to develop subtle or even obvious signs and symptoms of underlying cardiovascular disease. Only in this way, one patient at a time, will safer motherhood be possible for all with reduction of risk for maternal morbidity and mortality.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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