Review Article

Postpartum aortic dissection

Shi-Min Yuan*

Department of Cardiothoracic Surgery, First Hospital of Putian, Teaching Hospital, Fujian Medical University, Putian, Fujian Province, China

Abstract

Postpartum aortic dissection is a rare but lethal event. Until now, only a limited number of cases have been reported, and a comprehensive literature review from 1988 to 2012 yielded 27 cases. Postpartum aortic dissection occurred between Day 1 and Day 42 after delivery, either vaginally or by cesarean section. Pregnancy alone without underlying contributing risk factors and Marfan syndrome were the two main risk factors for postpartum aortic dissection, accounting for 44.4% and 40.7% of cases, respectively. Late presentation and delayed diagnosis may lead to sudden death. Improving prenatal and peripartum care is, therefore, crucial in preventing the development of aortic dissection. Prompt diagnosis and timely treatment of postpartum aortic dissection may prevent patient death.
Copyright © 2013, Taiwan Association of Obstetrics & Gynecology. Published by Elsevier Taiwan LLC. All rights reserved.

Keywords: bicuspid aortic valve; cardiac surgery; Marfan syndrome; pregnancy complications; sudden cardiac death

Introduction

Aortic dissection is a rare but potentially fatal event. Acute aortic dissection developing in association with pregnancy is even rarer, occurring during labor and delivery or in the early postpartum period [1,2]. Most aortic dissections occur in patients older than 50 years as a result of systemic hypertension and occur approximately 2−3 times more often in men [3]. However, women younger than 40 years of age may comprise half of the cases of aortic dissection in association with pregnancy and have neither risk factors nor typical clinical manifestations [4,5]. Among women, half of type B dissections occur during the postpartum period [6]. During pregnancy, aortic and vascular wall structures tend to be weaker and more susceptible to injury imposed by hemodynamic forces [7]. Fragmentation of the reticulum fibers, decreases in acid mucopolysaccharides, and loss of the normal corrugation of elastic fibers have been observed in the aortic walls of pregnant women [8]. The risk of aortic dissection or rupture appears to increase as gestation advances and persists for several months postpartum due to adaptational cardiovascular and hormonal changes occurring during and after pregnancy [9]. Women with an aortic root diameter > 40 mm, rapid dilation of aortic dimensions, and/or previous dissection of the ascending aorta are considered to be at high risk for aortic dissection [9], especially women with Marfan syndrome [10]. Until now, the emphasis has been on connective tissue disorders with regard to aortic dissection in pregnancy, but patients with no underlying contributing factors for its pathogenesis have been overlooked. The latter can be at more risk because unexpected aortic dissection may result in sudden death because of a delay in diagnosis and subsequent treatment [11]. Therefore, this medical emergency warrants further research.

Materials and methods

Literature search methods

The English-language literature on postpartum aortic dissection was comprehensively reviewed from the Medline database for 1988−2012. Additional searches were conducted

* Department of Cardiothoracic Surgery, First Hospital of Putian, Teaching Hospital, Fujian Medical University, 389 Longdejing Street, Chengxiang District, Putian 351100, Fujian Province, China.
E-mail address: shi_min_yuan@yahoo.com.

using the Google search engine. Retrieved articles were carefully reviewed and patient information was collected and analyzed.

Definitions

The postpartum period was defined as Days 1–42 after delivery. For acute aortic dissection, late presentation was defined as >2 hours from symptom onset, and early presentation was <2 hours from symptom onset. According to Harris et al [12], for patients with acute aortic dissection, the mean time from arrival at the emergency department to diagnosis was 4.3 hours (range: 1.5–24 hours) and from diagnosis to surgery was 4.3 hours (range: 2.4–24 hours). Therefore, a time from the patient's arrival at the emergency department to a diagnosis of >4 hours was regarded as a late/delayed diagnosis, and a time from diagnosis of aortic dissection to aortic surgery of >4 hours was considered as a delayed operation.

Statistical analysis

Quantitative data were expressed as mean ± standard deviation along with range and median. Comparisons of frequencies of events were done using Fisher's exact test. Probability–probability plot by conversion to natural logarithm was utilized for normal distribution examinations. Multiple logistic regression was used for predictive evaluation of patient survival/mortality. A p value < 0.05 was considered statistically significant.

Results

A total of 27 cases of aortic dissection occurring during the postpartum period were identified from 21 reports [3–7,13–28]. The mean patient age was 31.9 ± 4.6 years (range: 22–39 years, median: 32 years). Previous pregnancy history was para 1.8 ± 1.8 (range: 0–5, median: 1.5; $n = 16$): para 0 in five (31.3%) patients; para 1 and 2 in three (18.8%) patients each; para 3 in two (12.53%) patients; para 4 in one (6.3%) patient; and para 5 in two (12.5%) patients ($p = 0.6050$, Fisher's exact test). Type of delivery was recorded in 14 patients: seven (50%) had vaginal delivery with three having full-term infants; six (42.9%) patients underwent cesarean section; and one (9.1%) patient experienced fetal demise at 30 weeks gestation.

The onset of postpartum aortic dissection was 9.3 ± 9.0 days (range: 1–42 days, median: 7 days). Patient age, number of previous pregnancies, and onset time of postpartum aortic dissection were all normally distributed (Figs. 1–3). Information on the type of symptoms present at the onset of the dissection was available for 20 patients. Pain was the most common symptom and occurred in 12 (60%) patients. Pain was in the back in seven (58.3%) patients, the chest in two (16.7%), the left arm, back and abdomen in two (16.7%), and in both the chest and abdomen in one (8.3%) patient. Eleven (91.7%) of the 12 patients with pain had acute sharp pain, and the remaining patient had mild transient pain. Circulatory
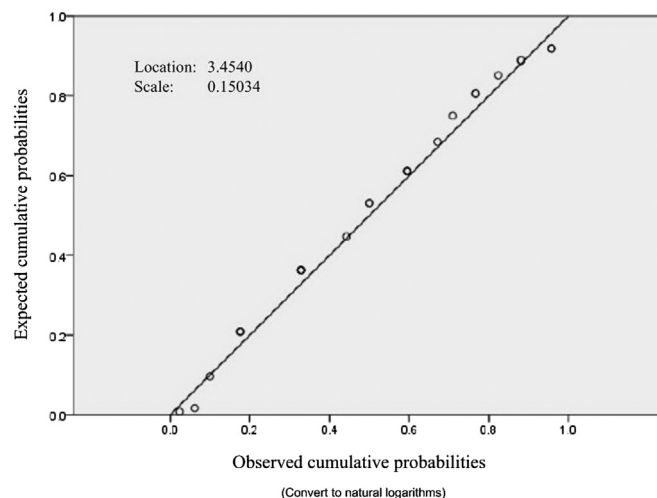

Fig. 1. Probability–probability plot of normal distribution of patients' age.

symptoms including dyspnea, shortness of breath, respiratory distress, or circulatory collapse were more common in these patients (Table 1). Three patients had a delayed presentation, arriving at the hospital 2 hours, 48 hours and 72 hours after the onset of aortic dissection, and the patient presenting at 72 hours died. A delayed diagnosis was made in two patients, at 96 hours and 144 hours after admission, and the first patient died. No patient in this cohort received delayed treatment.

There were two main predisposing risk factors: pregnancy with no underlying contributing factors and Marfan syndrome. These two risk factors were present in similar percentages of patients with aortic dissection (44.4% and 40.7%, respectively), with a significant difference when compared with other risk factors such as acute myocardial infarction, medial dystrophy (Erdheim's disease), bicuspid aortic valve, tobacco use, and obesity (Table 2).

Diagnostic methods used leading to a definite diagnosis were described for 20 patients. Computed tomography was the most commonly used diagnostic tool, followed by echocardiography. One patient died suddenly after acute onset of symptoms, and the diagnosis was made at autopsy (Table 3).
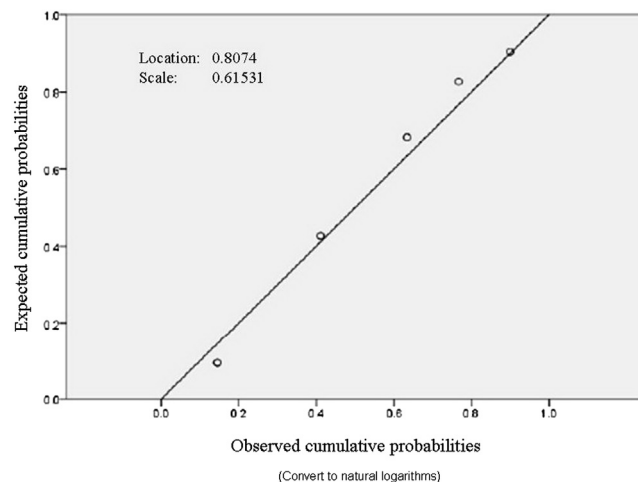

Fig. 2. Probability–probability plot of normal distribution of previous pregnancies.
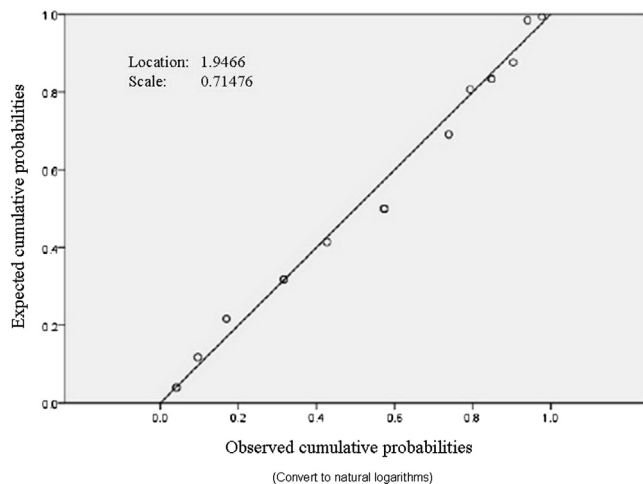
Fig. 3. Probability–probability of normal distribution of onset time of postpartum aortic dissection.

Sixteen (59.3%) patients had aortic dissection type A (one patient initially had type A which was followed by a type B aortic dissection 16 hours later), and type B was seen in 11 (40.7%) patients ($p = 0.2763$, Fisher's exact test). Aortic valve insufficiency was associated with aortic dissection in nine (34.6%) patients; the valvular insufficiency was mild in two (22.2%) and severe in seven (77.8%) patients ($p = 0.0567$, Fisher's exact test). In addition, severe mitral valve regurgitation was present in one patient. Massive pericardial effusions were present in two patients, pleural effusions in three, and both pericardial and pleural effusions in one.

Of the 27 patients, management of aortic dissection was surgical in 16 (59.3%), interventional in three (11.1%), conservative in four (14.8%), and initially conservative with eventual surgery in two (7.4%). However, two (15.4%) patients died suddenly after the onset of aortic dissection before treatment was possible (Table 4). Only Marfan syndrome and pregnancy with no additional contributing risk factors were present in six (37.5%) patients each, and cystic medial necrosis, tobacco use plus bicuspid aortic valve, obesity, and medial dystrophy (Erdheim's disease) were present in one

Table 1
Onset symptoms of aortic dissection in 20 pregnant patients.

Onset symptom	n (%)
Pain	12 (60)
Back pain	7 (35)
Chest pain	2 (10)
Left arm, back and abdomen	2 (10)
Chest, abdomen	1 (5)
Circulatory	7 (35)
Dyspnea, left leg hypothesia	2 (10)
Shortness of breath	3 (15)
Respiratory distress	1 (5)
Circulatory collapse	1 (5)
Asymptomatic	1 (5)
On observation of aortic dilation	1 (5)

Table 2
Risk factors responsible for the development of aortic dissection.

Risk factor	n (%)	p (Fisher's exact test)
Nil	12 (44.4)	
Marfan's syndrome (1 patient was associated with bicuspid aortic valve, s/p Yacoub procedure)	11 (40.7)	
Acute myocardial infarction	1 (3.7)	<0.0001
Medial dystrophy (Erdheim's disease)	1 (3.7)	
Bicuspid aortic valve, tobacco use	1 (3.7)	
Obesity	1 (3.7)	

(6.3%) patient each with type A aortic dissection. Marfan syndrome and pregnancy with no other contributing risk factors were present in five (45.5%) and six (54.4%) patients with type B aortic dissection. There were no significant differences regarding the predisposing risk factors (9.1% vs. 8.3%, $p = 1.0000$), between types A and B aortic dissections ($p = 1.0000$, Fisher's exact test). Type A aortic dissection was managed surgically in 14 (93.3%) patients, and no treatment was used in one (6.7%) patient who died suddenly. Type B aortic dissection was managed surgically in two (18.2%) patients, interventionally in three (27.3%), conservatively in four (36.4%), and conservatively followed by surgery in two (18.2%) patients. Significant differences were found in treatment strategies between Types A and B aortic dissection ($p < 0.0001$, Fisher's exact test).

Twenty-three (85.2%) patients survived and four (14.8%) died. The causes of death in the four women were sudden death in two and uncontrolled hypertension and postoperative complication in one each.

No significant differences were noted in mortality between Marfan patients and those with no other contributing risk factors (9.1% vs. 8.3%, $p = 1.0000$), for dissection Types A and B (25% vs. 0%, $p = 0.2693$), or for vaginal delivery and cesarean section (14.3% vs. 16.7%, $p = 1.0000$, Fisher's exact tests). There were no deaths among patients receiving interventional, conservative, or conservative/surgical treatments. A significant difference was found in mortality between patients receiving urgent aortic operation and those without an opportunity to be treated (12.5% vs. 100%, $p = 0.0500$, Fisher's exact test). Multiple logistic regression did not reveal any significant relationships between the above dependent variables and patient mortality ($\chi^2 = 6.195$, $p = 0.1025$ for overall model fit; and $p = 0.9986$ for mode of delivery, $p = 0.6240$ for

Table 3
Diagnostic means of aortic dissection in 20 patients.

Diagnostic means	n (%)
Computed tomography	9 (45)
Transthoracic echocardiography	4 (20)
Computed tomography + transesophageal echocardiography	2 (10)
Computed tomographic angiogram	2 (10)
Magnetic resonance imaging + transesophageal echocardiography	1 (5)
Pyelogram	1 (5)
Autopsy	1 (5)

Table 4
Management of aortic dissection.

Management	n (%)
Surgical operation	16 (59.3)
Bentall operation	3 (11.1)
Ascending aorta replacement + coronary ostium replacement/repair	3 (11.1)
Cabrol II operation	1 (3.7)
Aortic valve replacement, aortic root repair, descending aorta replacement	1 (3.7)
Ascending aorta–abdominal aorta bypass	1 (3.7)
Aortic arch repair + right and non-coronary ostium reconstruction + left coronary ostium reimplant	1 (3.7)
Atrial septal defect repair, aortic root + arch replacement, valve commissural resuspension, native right coronary ostium ligation, coronary artery bypass: saphenous vein graft to right coronary artery	1 (3.7)
Aortic valve repair, non-coronary ostium replacement, other 2 coronary ostia sparing, distal aorta replacement	1 (3.7)
Root replacement, aortic valve resuspension, coronary artery bypass: saphenous vein graft to right coronary artery	1 (3.7)
Aortic valve replacement, aortic root plication, descending aorta replacement	1 (3.7)
Stent graft	3 (11.1)
Conservative	4 (14.8)
Conservative and eventually surgery	2 (7.4)
No treatment	2 (7.4)

predisposing risk factor, and 0.9984 for type of aortic dissection).

Pathological study of the surgical aortic specimens was undertaken in nine patients; cystic medial degeneration was found in four (44.4%) patients, mucopolysaccharide deposit, diffuse mucoid imbibitions, medial dystrophy (Erdheim's disease), diffusely damaged tunica media, and myxoid changes were seen in one (11.1%) patient each. Three (11.1%) women had aortic dissection progression at 9–18 months follow-up and eventually required surgery.

Discussion

Postpartum aortic dissection is a rare event. The most common symptoms at onset were acute pains in the chest, back, abdomen, or left arm, followed by cardiopulmonary manifestations including shortness of breath, dyspnea, or circulatory collapse, and even sudden death. As previously described [9], such patients usually have larger aortic dimensions; however, aortic dimensions were not recorded in most patients described in this article.

The risk factors leading to aortic dissection can be multifactorial. Pregnancy-related hemodynamic stresses and hormonal effects are well recognized as the main risk factors responsible for the development of peripartum aortic dissection [4]. Hemodynamic stresses are significant in pregnant women in the second and third trimesters. These stresses include increased heart rate, stroke volume, cardiac output, left ventricular wall mass, and left ventricular end-

diastolic dimensions, and decreased systemic vascular resistance [29], heart rhythm changes [30], and compression to the abdominal vessels by the gravid uterus [31]. Pregnancy-induced alterations in plasma estrogen and progesterone concentrations are associated with aortic structural changes that are vulnerable to aortic dissection [32]. Coexisting risk factors include inherent disorders such as aortic diameter > 40 mm prior to conception [33], connective tissue disorders (Marfan, Turner, Loeys-Dietz, and Ehlers-Danlos syndromes and familial thoracic aorta aneurysm), bicuspid aortic valve, coarctation of the aorta, acute myocardial infarction, and obesity. Drug abuse can be another risk factor that may impose hemodynamic stress on the aortic wall and eventually lead to aortic dissection [34]. As a result of its rarity, postpartum aortic dissection may be associated with late presentation, delayed diagnosis, or misdiagnosis [3]. Knowledge of the patient's past medical history, including connective tissue disorders, bicuspid aortic valve, coarctation of the aorta, prior aortic surgery, or intravenous drug abuse, may predispose the physician to a high index of suspicion in diagnosing postpartum aortic dissection. Early presentation and a timely diagnosis with prompt treatment are crucial in preventing patient death.

Immer et al [2] reported 50 cases of puerpartum aortic dissection during a 20-year period; 40 were Type A aortic dissections and 10 were Type B. Five (12.5%) Type A and four (40%) Type B dissections occurred postpartum. One (20%) patient with Type A postpartum aortic dissection died. Similarly, our study showed a mortality of postpartum Type A aortic dissection of 26.7%, whereas no deaths occurred with Type B. More patients with Type A aortic dissection required aortic surgery than those with Type B.

Peripartum evaluation of the safety of an anticipated pregnancy should be carefully carried out prior to conception, labor, and vaginal delivery. In the event of connective tissue disorder, patients need to be under close surveillance with well-controlled peripartum hemodynamics. Moreover, it is essential to monitor the aortic dimensions during peripartum, because aortic dissection may occur at any time postpartum, varying from days to weeks [7]. Aortic root surgery should be done a few days after delivery for Type A dissection, and close monitoring and administration of β-blocking drugs should be continued for up to 3 months postpartum for both Types A and B aortic dissections [2]. Surgery for bicuspid aortic valve and coarctation of the aorta can be performed at any time during pregnancy when necessary [35,36].

From the present study, patients with postpartum aortic dissection were young, with an age range of 22–39 years, which is in agreement with what has been reported in the literature [4,5]. It was notable that postpartum aortic dissection may develop with no predisposing factors regarding patients aged 22–29 years, para number 0–5, and onset time of 1–42 days post-delivery by either vaginal delivery or cesarean section. The two predisposing risk factors of pregnancy alone with no underlying contributing factors and Marfan syndrome prevailed as the leading causes for the development of aortic dissection, with significant differences when compared to

other risk factors. In the present study, late presentation and delayed diagnosis caused death in 33.3% and 50% of patients.

In conclusion, postpartum aortic dissection is rare. It may occur without predilections regarding the postpartum period, patient age during pregnancy, and prior pregnancy history. Late presentation and delayed diagnosis may cause death in these patients. Therefore, a suspicion of aortic dissection should arise when pregnant women present with complaints of acute pain in the early postpartum period, in those with connective tissue disorder, or even in those with no additional risk factors.

References

[1] Braverman AC. Acute aortic dissection: clinician update. Circulation 2010;122:184–8.

[2] Immer FF, Bansi AG, Immer-Bansi AS, McDougall J, Zehr KJ, Schaff HV, et al. Aortic dissection in pregnancy: analysis of risk factors and outcome. Ann Thorac Surg 2003;76:309–14.

[3] Kang BH, Lee MA, Rhee YE, Noh HT. Unexpected acute aortic dissection after elective cesarean section delivery: report of a case and review of the literature. Korean J Obstet Gynecol 2011;54:696–700.

[4] Gelpi G, Pettinari M, Lemma M, Mangini A, Vanelli P, Antona C. Should pregnancy be considered a risk factor for aortic dissection? Two cases of acute aortic dissection following cesarean section in non-Marfan nor bicuspid aortic valve patients. J Cardiovasc Surg (Torino) 2008;49:389–91.

[5] Petrillo JM, Mendez O, Vakiener A, Back E. Postpartum aortic dissection: a case report and brief literature review. South Med J 2004;97:S12–3.

[6] Rosenberger LH, Adams JD, Kern JA, Tracci MC, Angle JF, Cherry KJ. Complicated postpartum type B aortic dissection and endovascular repair. Obstet Gynecol 2012;119:480–3.

[7] Gandhi SD, Iqbal Z, Markan S, Almassi GH, Pagel PS. Massive retrograde acute type B aortic dissection in a postpartum woman with a family history of Marfan syndrome. J Clin Anesth 2008;20:50–3.

[8] Master M, Day G. Acute aortic dissection in pregnancy in a woman with undiagnosed Marfan syndrome. Case Rep Obstet Gynecol 2012. Article ID 490169, http://dx.doi.org/10.1155/2012/490169.

[9] Silversides C, Colman J, Kennie A, Balint OH, Bottega N, Burchill L, et al. Heart disease and pregnancy: Marfan syndrome. http://www. heartdiseaseandpregnancy.com/pdf/phy_mar.pdf [accessed 10.08.13].

[10] Vranes M, Velinovic M, Kovacevic-Kostic N, Savic D, Nikolic D, Karan R. Pregnancy-related aortic aneurysm and dissection in patients with Marfan's syndrome: medical and surgical management during pregnancy and after delivery. Medicina (Kaunas) 2011;47:604–6.

[11] Braverman AC. Aortic dissection: prompt diagnosis and emergency treatment are critical. Cleve Clin J Med 2011;78:685–96.

[12] Harris KM, Strauss CE, Eagle KA, Hirsch AT, Isselbacher EM, Tsai TT, et al. Correlates of delayed recognition and treatment of acute type A aortic dissection: the International Registry of Acute Aortic Dissection (IRAD). Circulation 2011;124:1911–8.

[13] Snir E, Levinsky L, Salomon J, Findler M, Levy MJ, Vidne BA. Dissecting aortic aneurysm in pregnant women without Marfan disease. Surg Gynecol Obstet 1988;167:463–5.

[14] Nolte JE, Rutherford RB, Nawaz S, Rosenberger A, Speers WC, Krupski WC. Arterial dissections associated with pregnancy. J Vasc Surg 1995;21:515–20.

[15] Scalia D, Rizzoli G, Scomparin MA, Venturini A, Casarotto D. Aortic dissection in puerperium: a case report. J Heart Valve Dis 1996;5:251–3.

[16] Lipscomb KJ, Smith JC, Clarke B, Donnai P, Harris R. Outcome of pregnancy in women with Marfan's syndrome. Br J Obstet Gynaecol 1997;104:201–6.

[17] Lind J, Wallenburg HC. The Marfan syndrome and pregnancy: a retrospective study in a Dutch population. Eur J Obstet Gynecol Reprod Biol 2001;98:28–35.

[18] Hsieh TH, Chao TH, Chang CJ, Chen JH. Acute aortic dissection associated with left ventricular dysfunction in a postpartum and normotensive young woman. J Formos Med Assoc 2003;102:331–3.

[19] Chavanon O, Rama A, Leprince P, Bonnet N, Pavie A, Jondeau G, et al. Valve-sparing operation in a young woman with Marfan syndrome: a word of caution. J Thorac Cardiovasc Surg 2006;132:683–4.

[20] Omar AR, Goh WP, Lim YT. Peripartum acute anterior ST segment elevation myocardial infarction: an uncommon presentation of acute aortic dissection. Ann Acad Med Singapore 2007;36:854–6.

[21] Radermecker MA, Durieux R, Canivet JL, Limet R. Metachronous type III and type II acute aortic dissections in puerperium. Eur J Cardiothorac Surg 2007;32:541–3.

[22] Savi C, Villa L, Civardi L, Condemi AM. Two consecutive cases of type A aortic dissection after delivery. Minerva Anestesiol 2007;73:381–3.

[23] Neelakandhan1 KS, Parvathy U, Gopakumar D, Geothe J. Descending aortic aneurysm with acute type III dissection in a postpartum patient. Ind J Thorac Cardiovasc Surg 2007;23:256–9.

[24] Bjørnstad H, Hovland A. Acute postpartum aortic dissection. Int J Gynaecol Obstet 2008;103:68–9.

[25] Pacini L, Digne F, Boumendil A, Muti C, Detaint D, Boileau C, et al. Maternal complication of pregnancy in Marfan syndrome. Int J Cardiol 2009;136:156–61.

[26] Nishino H, Suda K, Kuramaoto A, Honda Y, Takemiya K, Ishii H, et al. Stanford type B aortic dissection associated with pregnancy in patients with Marfan syndrome – a case report and review of the literature. J Cardiol Cases 2010;1:e180–3.

[27] Nishad AAN, Herath R, Silva GRC, Mohommad R, Wijesinghe P, Padumadasa S, et al. A fatal case of dissecting aortic root aneurysm due to undiagnosed Marfan syndrome in the puerperium. Sri Lanka J Obstet Gynaecol 2012;34:55–7.

[28] Huang J, Liu H, Ding YL. Two cases of acute aortic dissection following preeclampsia in non-Marfan patients. Chin Med J (Engl) 2012;125:2073–5.

[29] Mesa A, Jessurun C, Hernandez A, Adam K, Brown D, Vaughn WK, et al. Left ventricular diastolic function in normal human pregnancy. Circulation 1999;99:511–7.

[30] Adamson DL, Nelson-Piercy C. Managing palpitations and arrhythmias during pregnancy. Heart 2007;93:1630–6.

[31] Katz VL, Hansen AR. Complications in the emergency transport of pregnant women. South Med J 1990;83:7–10.

[32] Landau R, Smiley RM. Anesthesia for cardiac surgery in the pregnant patient. http://tele.med.ru/book/cardiac_anesthesia/text/es/es029.htm [accessed 10.08.13].

[33] Meijboom LJ, Vos FE, Timmermans J, Boers GH, Zwinderman AH, Mulder BJ. Pregnancy and aortic root growth in the Marfan syndrome: a prospective study. Eur Heart J 2005;26:914–20.

[34] Eagle KA, Isselbacher EM, DeSanctis RW. International Registry for Aortic Dissection (IRAD) Investigators. Cocaine-related aortic dissection in perspective. Circulation 2002;105:1529–30.

[35] Datt V, Tempe DK, Virmani S, Datta D, Garg M, Banerjee A, et al. Anesthetic management for emergency cesarean section and aortic valve replacement in a parturient with severe bicuspid aortic valve stenosis and congestive heart failure. Ann Card Anaesth 2010;13:64–8.

[36] Plunkett MD, Bond LM, Geiss DM. Staged repair of acute type I aortic dissection and coarctation in pregnancy. Ann Thorac Surg 2000;69:1945–7.