Anesthetic Challenges for Modified Bentall Procedure in a Pregnant Marfan Patient with Acute Stanford Type A Dissection

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Abstract

Aortic dissection is a life-threatening condition which requires immediate surgical intervention. It has been estimated that half of aortic dissection and/or ruptures in women younger than 40 years of age have been associated with pregnancy.[19] The limited data on the coincidence of Marfan syndrome, pregnancy and aortic dissection makes its anesthetic management a formidable challenge to any anesthesiologist.

This is a case of a 33-year-old G1P0, 28 weeks age of gestation with Marfan syndrome, who had Aortic Dissection Stanford type A and underwent an emergency Modified Bentall's surgery with cardiopulmonary bypass and deep hypothermic circulatory arrest.

This case illustrates the dilemma of managing this case since there are two patients involved, mother and fetus. Wrong decision could result in demise of both. Although the aim is life for both, survival of one without neurologic sequelae is already considered a success.

Keywords: Marfan syndrome, Modified Bentall Procedure, Pregnancy, Acute type A dissection

Abstrak

Diseksi aorta adalah kondisi yang mengancam jiwa yang memerlukan intervensi bedah segera. Diperkirakan bahwa setengah kasus dari diseksi aorta dan / atau ruptur aorta pada wanita yang lebih muda dari 40 tahun berhubungan erat dengan kehamilan. 19 Keterbatasan data mengenai manajemen anestesi pada pasien hamil dengan sindrom Marfan yang mengalami diseksi aorta merupakan tantangan tersendiri pada seorang ahli anestesi .

Berikut ini adalah laporan kasus seorang pasien hamil G1P0 berusia 33 tahun, usia kehamilan 28 minggu dengan sindrom Marfan, yang memiliki Diseksi aorta Stanford tipe A dan menjalani operasi darurat Bentall's Modifikasi dengan *bypass* kardiopulmonari dan *deep hypothermic circulatory arrest*. Laporan ini menggambarkan dilema yang timbul dalam pengelolaan kasus dimana terdapat dua pasien yang terlibat, ibu dan janin. Keputusan yang salah bisa mengakibatkan kematian keduanya. Meskipun tujuannya adalah untuk kelangsungan hidup ibu dan janin, kelangsungan hidup salah satu pasien tanpa gejala sisa neurologis merupakan suatu keberhasilan.

Kata kunci : sindroma marfan, Prosedur Bentall, Kehamilan, Diseksi aorta tipe A akut

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Introduction

Acute aortic dissection of the ascending aorta (Stanford type A) is highly lethal. Forty percent of patients who reach the emergency department die immediately. This mortality rate is increased by 1% per hour in the first 48 hours and between 5%-20% die during or shortly after the surgery.[2,24,32]

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Early death may occur as a result of malperfusion syndromes (cerebrovascular, visceral, renal, or peripheral ischemia), cardiac complications (acute aortic insufficiency, coronary ischemia, cardiac tamponade), or free rupture.[17] Surgery, which aims to prevent aortic rupture, cardiac complications and other end-organs damage, is the definitive treatment for patient with type A acute aortic dissection.

Deep hypothermic circulatory arrest (DHCA) is technique required in cases of acute aortic dissection or aneurysms that extend into the aortic arch.[49]

Surgical replacement of the aortic valve and surgery of the aortic aneurysm or dissection that extend into the aortic arch carry risk of neurologic injury from global ischemia or embolization of atherosclerotic debris secondary to clamping of the great vessels.[5] Surgery of descending aorta is associated with postoperative paraplegia secondary to interruption of 13 to 17% of blood supply of the spinal cord.[5]

This case report will show how neurologic sequelae was prevented during anesthesia for modified Bentall procedure to a pregnant Marfan patient with acute type A dissection.

Case summary

This is a case of a 33-year-old primigravid, 28 weeks age of gestation (AOG) with Marfan syndrome, who consulted due to sudden chest discomfort with radiation to the left jaw associated with burning substernal chest pain and diaphoresis.

Past medical history showed that the patient had hiatal hernia and gastroesophageal reflux disease (GERD) and was on esomeprazole tab as needed. She had no previous surgery. Prenatal check up was not clear.

Family history showed that her mother and brother have Marfanoid features. Physical examination revealed a conscious, coherent patient who arrived via stretcher. She weighs 60 kg and stands160 cm. Her vital signs were as follows: blood pressure (BP) 106/69 mmHg on both upper extremities, 110/71 on both lower extremities, heart rate (HR) 97-100 bpm, respiratory rate (RR) 20 per minute. Airway examination revealed mallampati I classification with good neck movement. Chest examination revealed a pectus carinatum deformity. Her abdomen was globular. Fetal heart tone was appreciated at 150 beats per minute. Cephalic presentation was palpable. No uterine contractions were noted. Internal examination revealed a soft and close cervix. Her arm span of 174 cm with positive wrist and thumb sign were noted. Hindfoot deformity was also observed. Good peripheral pulses were appreciated. Neurologic examination revealed intact cranial nerves, normal motor strength, no tingling nor numbness, normal response of tendon reflexes with no pathologic reflexes found.

Diagnostic workups revealed anemia with leucocytosis, neutrocytosis and hypocalcemia (see Appendix A). Carotid duplex scan showed > 50% stenosis in the left common carotid artery. Positive troponin I indicates myocardial necrosis. Transesophageal echocardiography (TEE) showed a hypocontractile free walls of the right ventricle. An echocardiography showed aortic dissection confirmed by magnetic resonance angiography (MRA) which showed aortic dissection Stanford type A (Fig. 1A & 1B).

Patient was diagnosed to have aortic aneurysm and dissection Stanford type A with carotid stenosis and right coronary artery disease, Marfan syndrome, G1P0 28 weeks AOG cephalic, not in labor. She was classified as ASA class III-E.

Modified Bentall's procedure with double set up for possible obstetrical intervention was contemplated. Risks and benefits were explained to patient and relatives. The risk of demise for both the patient and fetus was not eliminated. Survival of patient but with neurologic and nephrologic sequelae was also a possibility. There was no immediate indication for an emergency caesarian section and fetal demise can occur. The risks were explained by all specialists — obstetrician, cardiologist, thoracic, cardiovascular surgeon, intensivist, neonatologist and anesthesiologist. Patient and relatives accepted risks. Urgent modified Bentall procedure was scheduled.

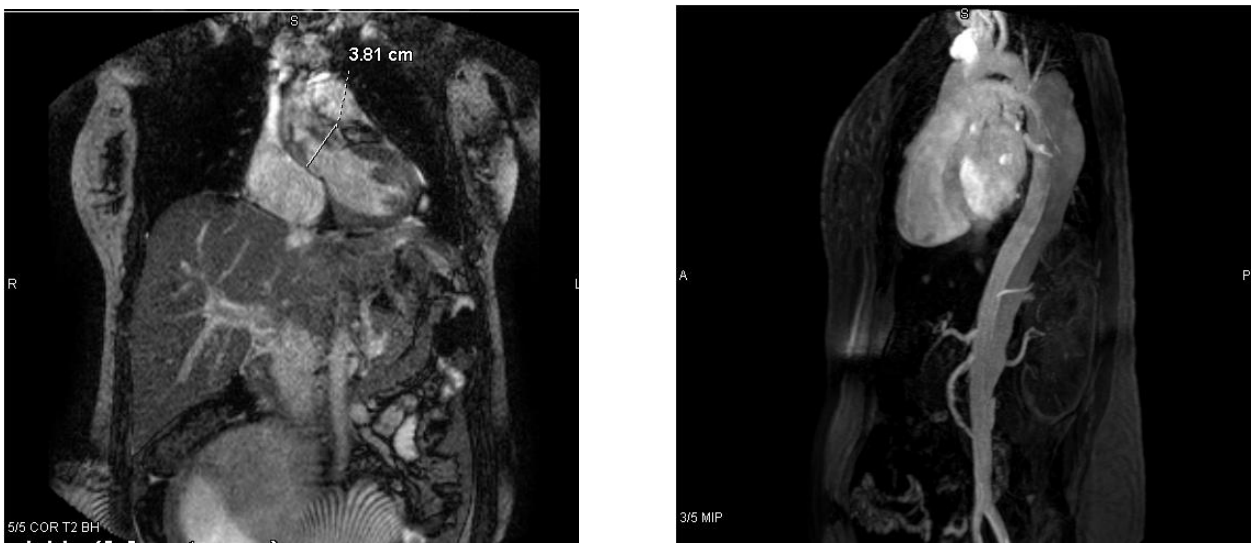

Figure 1A&B. MRA of the chest/thoracic (9/29/10) showed aortic aneurysm with dissection at the level of proximal ascending aorta down to the visualized common iliac arteries.

Patient was given betamethasone 12 mg IM and cefuroxime IV prior to the surgery. Packed RBC, thawed fresh frozen plasma, platelet concentrate and fresh whole blood were prepared for use Monitors placed were as follows: temperature probe on the nasopharynx, 5 leads-ECG, end tidal CO2, arterial line at right radial artery and left brachial artery, and pulmonary artery catheter inserted in the right internal jugular vein. Urethral catheter was inserted to monitor the urine output hourly.

Initial vital signs were as follows: BP 165/60 mmHg, HR 105 bpm and pulmonary arterial pressure (PAP) 25/19 mmHg (normal PAP is 15-28/ 5-16mm Hg).[52]

Initial arterial blood gas showed a metabolic acidosis (see Appendix A) and was corrected with the NaHCO3 administration. Nitroglycerin drip was started during induction period to achieve hemodynamic stability during intubation and to improve coronary blood flow.

Anesthetics administered were as follows: fentanyl 250µg followed by infusion, midazolam 1 mg, propofol infusion, rocuronium 50 mg followed by infusion and sevoflurane titrated between 1-3%. Patient was intubated with 7.5 size ETT under direct visualization of vocal cords. She was hooked to ventilator with the setting as follows: volume controlled ventilation, tidal volume 450 ml, RR 14 per minute, IE ratio 1:2 and FiO2 1.

during surgery. Proton pump inhibitor and dexamethasone 15 mg iv in a gauge 16 peripheral IV line were administered during preinduction period.

Transesophageal echocardiography (TEE) done after induction showed a dilated aortic root of 44 mm x 38 mm (normal < 37mm)[41] and proximal ascending aorta 35 mm x 27 mm (normal is 30 mm)[21] with dissection of the posterior aortic wall from the aortic root to the descending thoracic aorta of 32 mm (normal 18 – 22 mm)[21] with thrombus formation at the false lumen. There was also moderate to severe aortic regurgitation with mild mitral and tricuspid regurgitation.

Citicholine 1 gm, tranexamic acid 2 g were administered. Nitroglicerine drip, low dose dopamine drip and blood transfusion were started. Median sternotomy commence 29 hours from onset of symptoms or five hours from arrival at the emergency department. Heparin at 3 mg/kg was administered. Activated coagulation time (ACT) level of 375 was achieved before cannulation of the axillary artery, femoral artery and right atrium. ACT determination was done every 30 minutes to maintain a level above 350.

Surgery proceeded with a period of cardiac arrest by infusion of cardioplegic solution into the aortic root. Deep hypothermia at 20°C was achieved by infusion of cold blood using CPB, application of blanket roll and ice pack over the head. After the core temperature of 20C was achieved, innominate artery was clamped.

Blood cardioplegic solution was given in retrograde manner to facilitate the aortic root replacement with a composite valve graft. During this period of time, antegrade cerebral perfusion technique was used to provide blood flow to the brain. Perfusion to other organs was also preserved through femoral artery cannulation Rewarming of the temperature to 28°C was then initiated and maintained during completion of the coronary artery bypass of the right main coronary artery using a distal saphenous vein graft.

All anesthetic agents were given by continuous intravenous infusion. Total DHCA time, clamped/ischemic time and CPB time were 24 minutes, 155 minutes and 190 minutes respectively.

After rewarming to 37˚C, the bypass was gradually terminated. Heparin reversal with protamine was administered intravenously.

The post cardiopulmonary bypass course was uneventful. The PAP was maintained at 17-30/15-20 mmHg, ejection fraction was 69% compared to 63% preoperatively.

The modified Bentall surgery lasted 7 hours. Total blood loss was 1000 cc (maximal allowable blood loss was 252 cc) with total urine output of 1350 cc. Blood products transfused were PRBC of 4 units, FFP of 3 units and platelet concentrate of 5 units.

She was maintained on mechanical ventilator for six hours at the post anesthesia care unit (PACU). Postoperative fetal heart tones could not be appreciated. Pelvic ultrasound showed fetal demise. Expectant management for the intrauterine fetal demise (IUFD) was the best option at this time.

Patient was transferred to coronary care unit sedated but arousable with pain controlled by tramadol infusion and later by oral combination of paracetamol 325 mg and tramadol 37.5 mg.

On 2nd post op day, left ventricular dysfunction was noted on 2D echo with dyskinesia on the entire interventricular septum and hypokinesia on the anterior left ventricle (LV) free wall with best contraction at the antrolateral and inferolateral LV free wall (EF 33% by Tiechholz and 49% by Simpson's).

Dobutamine drip 5-15 mcg/kg/min was started and titrated. Fondaparinux 2.5 mg was also administered subcutaneously as an antithromboembolic agent. Other management includes general liquid diet progressive heart delight, humulin R sliding scale, calcium 600 mg, Vit D 400 IU tablet p.o. and potassium chloride tab 750 mg p.o. Neurologic examinations revealed same as preinduction.

Repeat 2D echo on 4th post op day showed an improvement in LV function (EF 51% by Simpson's) and wall motion (hypokinesia of entire septum).

She stayed at the coronary care unit for 6 days. No evidence of neurologic, nephrologic deficit, pulmonary and cardiac problems was noted. . Patient was discharged with improvement and preferred to be transferred to another hospital for delivery of the fetus.

Discussion

We are presented with a patient with aortic aneurysm with dissection type A. This requires immediate surgical intervention because of 50% risk of rupture.[2,24,32]

The presence of aortic dissection and systemic features with score ≥7 made our patient fulfill criteria of having Marfan syndrome following the 2010 Revised Ghent nosology criteria (see Appendix B). The aortic dissection in Marfan's syndrome patient usually manifests during pregnancy which happened in this case.

Type A dissection, meaning dissection involving the ascending aorta (Fig. 2) present in the patient, carries the risk of neurologic sequelae when the aorta is clamped.

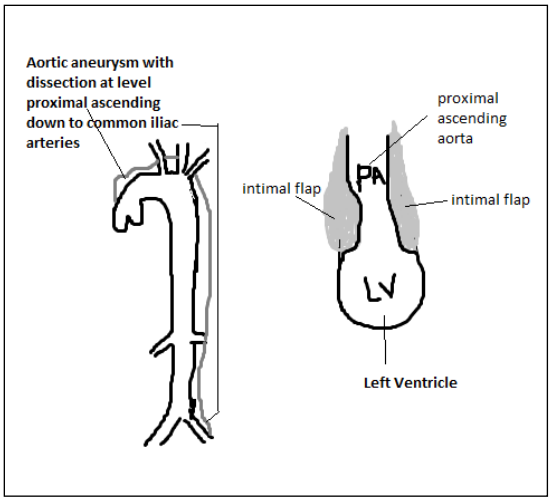

Figure 2. Type A aortic aneurysm and dissection at the level of the proximal ascending down to the common iliac arteries (left) and intimal flap from proximal ascending aorta (right)

In addition, the presence of more than 50 % stenosis of the left carotid artery, though not yet an indication for carotid endarterectomy, also added to the risk of cerebral ischemia. Thus, cardiopulmonary bypass and deep hypothermic circulatory arrest techniques, defined as the use of systemic hypothermia (core temperature of 14-20°C)[3] and the intentional cessation of the circulation for periods up to 60 minutes[30], were employed by placing the patient at 20°C to reduce the cerebral metabolic rate and oxygen consumption, and thus minimize the complication of cerebral ischemia.

Recurrence of aortic dissection in this patient is likely to happen especially during subsequent pregnancy.[19,23] Thus, grafting of the aortic dissection will not be enough. There should also be an aortic valve replacement and reimplantation of the left coronary ostium with a long interposed graft wrapping behind the grafting of the aortic dissection.[38]

The type A aortic dissection which also disrupted the right coronary artery blood flow (Fig. 3) required the patient to undergo coronary artery bypass graft in addition to the aortic dissection surgery and aortic valve replacement. Thus, the extension of the duration of the surgery is unavoidable.

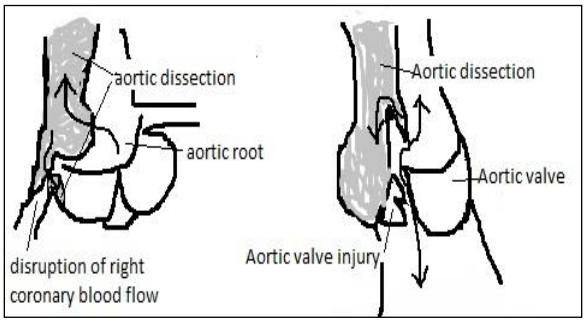

Figure 3. Disruption of the right coronary blood flow (left) and injury to the aortic valve due to aortic dissection causing moderate-severe aortic regurgitation (right)

However, the problems did not end there. The patient was also a primi gravid at 28 weeks AOG. Her prenatal check up and work-ups were not available. Referral to the Obstetric department could not find any indication for elective caesarian section at that time. The patient was not in labor. The fetal lung was expected to be immature as surfactant would be released at 30 weeks AOG, at which time could be detected in amniotic fluid.[42] Forty eight hours of steroids (dexamethasone every 12 hours for 4 doses or bethametasone every 24 hours for 2 doses) should be administered to accelerate fetal lung maturation.[11] However, the emergent nature of the surgical condition could not afford to wait 48 hours. Aspiration of amniotic fluid to detect the presence of surfactant at 28 weeks AOG seemed to have no beneficial effect.[42]

If elective caesarian section was done in a preterm patient who is not in labor and with very low birth weight fetus with immature lung, the chance of survival of the fetus would be less than 50%.[10] Bleeding could be expected from the uterus during the caesarean section in full term pregnancy with blood loss as much as 500 ml – 1000 ml in 75% of women having a caesarean section in the Philippines and more than 1000 ml in 4% of that population.[18] Furthermore, the life of the post partum patient would be greatly endangered if the patient would undergo aortic surgery wherein anticoagulants would be administered.

The patient's condition would have more than 45% risk of mortality if not immediately operated.[24] An increase of 1-2% mortality was expected for every hour delay of surgery from onset of symptoms.[24]

The risks and benefits were explained to the patient and the relatives. The family and the physicians agreed to do the aortic surgery to save the life of the pregnant patient without any neurologic sequelae with the hope that the fetus would be able to survive the insult caused by the medical and surgical conditions.

The worst possible scenarios were also explained to the relatives. The worst possibility was that the patient and the fetus might not be able to survive the surgery. Another possibility was the patient might survive the surgery but with neurologic sequelae or with kidney problem or with lower extremity paralysis secondary to the clamping of the aorta, carotid stenosis and prolonged surgery. Yet, the more expected possibility was the patient might survive but the fetus might not.

Since the patient was brought to the emergency department 24 hours from onset of symptoms, the patient was immediately operated five hours after admission or 29 hours after onset of symptoms.
TEE was done after induction of GEA to determine the final surgery. Ascending aortic replacement, total arch replacement with reattachment of brachiocephalic branches with modified Bentall procedure (the replacement the aortic root and reimplantation the left coronary artery) were chosen to minimize the risk of the recurrence of the later-onset aneurysms and dissection of the aorta which commonly encountered in Marfan syndrome.[23]

The modified Bentall technique, which was reported by Svensson in 1992, is an aortic root's replacement using a composite valve graft (bileaflet mechanical valve attached to a polyester tube graft) involving a left coronary ostium's reimplantation using Kochoucous's button modification by leaving button's of aortic wall surrounding both coronary arteries, which are then mobilized and sutured to the aortic graft (Fig 4).[38, 48]
Standard cardiopulmonary bypass (heart lung machine) and deep hypothermic circulatory arrest were achieved to facilitate the aortic dissection repair, valve replacement and CABG. Usual cannulation was impossible to use due to the location of the aortic dissection, thus axillary and femoral arteries were used for site of cannulation to maintain blood flow to the brain and other organs.

The patient was extubated six hours post surgery without any neurologic sequelae.

Figure 4. Illustration of the modified Bentall procedure for aortic root replacement, during which buttons of aortic wall surrounding coronary arteries are mobilized and sutured to the aortic graft. (Reprinted from Schwartz's Principles of surgery, 8th edition, 2004)

However, fetal heart tone could no longer be appreciated post operatively. Thus, though the difficult CABG and aortic surgery can be considered a success, the demise of the fetus intrautero made its success not in toto.

Aware of the all the consequences that might occur, the patient and the relatives appreciated the effort of the team who was able to prevent the worst possibilities that might occur to the patient.

The anesthetic management for modified bentall procedure in a pregnant patient with Acute Type A dissection is very challenging since there are two patients involved, mother and child. The additional risk for higher maternal morbidity which related to Marfan syndrome also made the nature of the event more challenging for any anesthesiologist.

Although the aim is life for both, survival of one without neurologic sequelae is already considered a success.

Conclusion

References

1. Akashi H, Tayama K, Fujino T, Onitsuka S, Sakashita H, Aoyagi S. Surgical treatment for acute type A aortic dissection in pregnancy: a case of aortic root replacement just after cesarean section. *Jpn Circ J*. 2000;64:729–730.

2. Anagnostopoulos CE, Prabhakar MJ, Kittle CF. Aortic dissections and dissecting aneurysms. Am J Cardiol. 1972;30:263–73.
3. Andropoulos D et. al. Anesthesia for congenital heart disease 2nd ed. 2010: Chap 2; p 168

4. Assad RS, Lee FY, Bergner K, Hanley FL. Extracorporeal circulation in the isolated in situ lamb placenta: hemodynamic characteristics. J Appl Physiol 1992;72:2176–80.

5. Barash P, et al. Clinical Anesthesia 6th edition, 2009 : 1073-1105

6. Baydin A, Nargis C, Nural MS, et al. Painless, acute aortic dissection `presenting as an acute stroke. Mt Sinai J Med. 2006;73:1129 –31.

7. Blackburn S. Maternal, Fetal & Neonatal Physiology. A Clinical perspective. 2007 : Chap 10. P 336

8. Chow SL. Acute aortic dissection in a patient with Marfan's syndrome complicated by gestational hypertension. Med J Aust. 1993 Dec 6-20;159(11-12):760-2.

9. Chirillo F, Salvador L, Bacchion F, et al. Clinical and anatomical characteristics of subtle-discrete dissection of the ascending aorta. Am J Cardiol. 2007;100:1314 –9.

10. Committee to Study the Prevention of Low Birthweight and Division of Health Promotion and Disease Prevention. Preventing Low birth weight. 1985: p 28

11. Creasy R et al. Maternal-fetal medicine:principles and practice.5th ed. 2004:p460

12. Datta S. Anesthetic and Obstetric management of high risk pregnancy: Cardiovascular disease in pregnancy. p166

13. Dietz HC, Pyeritz RE. Mutations in the human gene for fibrillin-1 (FBN1) in the Marfan syndrome and related disorders. Hum Mol Genet.1995;4 Spec No:1799–809.

14. Elkayam U, Gleicher N. Hemodynamics and cardiac function during normal pregnancy and the puerperium. In: Elkayam U, Gleicher N, eds. *Cardiac Problems in Pregnancy*.New York, NY: Wiley-Liss; 1998:23–32.

15. Elkayam U, Ostrzega E, Shotan A, Mehra A. Cardiovascular problems in pregnant women with the Marfan syndrome. Ann Intern Med 1995;123:117–22

16. Espinoza S C, Selman A R, Pauchard T F, Rivera F J, Iturra U S, Montecinos R F, Eliash D H, Undurraga H F. Acute type A aortic dissection in pregnant patient with Marfan syndrome. Report of one case. Rev Med Chil. 2009 Jan;137(1):98-100. Epub 2009 Apr 23. Spanish

17. Estrera AL, Huynh TT, Porat EE, et al: Is acute type A aortic dissection a true surgical emergency?. *Semin Vasc Surg* 2002; 15:75-82.

18. Festin et al. Caesarean section in four South East Asian countries: reason for, rates, associated care practices and health outcomes. BMC Pregnancy childbirth v. 9. 2009: 9-17

19. Goland S, Elkayan U. Cardiovascular Problem in Pregnant Women with Marfan Syndrome. Circulation 2009;119;619-623

20. Gott VL, Cameron DE, Alejo DE, Greene PS, Shake JG, Caparrelli DJ, Dietz HC. Aortic root replacement in 271 Marfan patients: a 24-year experience. *Ann Thorac Surg*. 2002;73:438–443

21. Harrison's Principles of Internal Medicine 16th edition, 2005 : pp 34, pp 2329-2330, p 1481

22. Hagan PG, Nienaber CA, Isselbacher EM,Bruckman D, Karavite DJ, Russman PL,Evangelista A, Fattori R, Suzuki T, Oh JK, Moore AG, Malouf JF, Pape LA, Gaca C, Sechtem U, Lenferink S, Deutsch HJ, Diedrichs H, Marcos y Robles J, Llovet A, Gilon D, Das SK, Armstrong WF, Deeb GM,Eagle KA. International Registry of Acute Aortic Dissection (IRAD): new insights from an old disease. *JAMA.* 2000;283:897–903.

23. Hiratzka et al. Guidelines for the Diagnosis and Management of Patients With Thoracic Aortic Disease. JACC Vol. 55, No. 14, 2010 :e27–129

24. Hirst Jr AE, Johns Jr VJ, Kime Jr SW: Dissecting aneurysm of the aorta: A review of 505 cases. *Medicine* 1958; 37:217-219.

25. Immer FF, Bansi AG, Immer-Bansi AS, McDougall J, Zehr KJ, Schaff HV, Carrel TP. Aortic dissection in pregnancy: analysis of risk factors and outcome. *Ann Thorac Surg*. 2003;76:309 –314.

26. Jondeau G, Nataf P, Belarbi A, Farcot JC, Iung B, Delorme G, Gandjbakhch I, Bourdarias JP. Aortic dissection at 6 months gestation in women with Marfan's syndrome: simultaneous Bentall intervention and cesarean section [in French]. *Arch Mal Coeur Vaiss*. 2000;93:185–187

27. Kirklin J, Barratt-Boyes et al. Hypothermia, Circulatory Arrest, and Cardiopulmonary Bypass. Cardiac surgery. New York: Churchill Livingstone; 1993:61-127

28. Lachandani S, Wingfield M. Pregnancy inwomen with Marfan's syndrome. *Eur J Obstet Gynecol*. 2003;110:125–130

29. Loeyz, et al. The revised Ghent nosology for the Marfan syndrome. J Med Genet 2010;47:476-485

30. Mackay J, Arrowsmith J. Core topics in cardiac anesthesia. 1st ed, 2004; p279

31. Maruyama Y, Oguma F, Kosuge T, Yokosawa T, Eguchi S. Successful repair of an acute type A dissection during pregnancy. Nippon Kyobu Geka Gakkai Zasshi. 1990 Nov;38(11):2296-9.

32. Masuda Y, Yamada Z, Morooka N, et al. Prognosis of patients with medically treated aortic dissections. Circulation. 1991;84:III7–13

33. Meijboom LJ, Drenthen W, Pieper PG, Groenink M, van der Post JA, Timmermans J, Voors AA, Roos-Hesselink JW, van Veldhuisen DJ, Mulder BJ. Obstetric complications in Marfan syndrome. *Int J Cardiol*. 2006;110:53–59.

34. Milewicz DM, Dietz HC, Miller DC. Treatment of aortic disease in patients with Marfan syndrome. Circulation. 2005;111:e150 –7.

35. Mul TF, van Herwerden LA, Cohen-Overbeck TE, Catsman-Berrevoets CE, Lotgering FK. Hypoxic-ischemic fetal insult resulting from maternal aortic root replacement, with normal fetal heart rate at term. Am J Obstet Gynecol 1998;179(3Pt 1):825–7.

36. Murray CA, Edwards JE. Spontaneous laceration of ascending aorta.Circulation. 1973;47:848 –58.

37. Naito H, Naito H, Tada K. Open heart operation for a pregnant patient with Marfan syndrome [in Japanese]. *Mastui*. 2005;54:525–529

38. Nakahira A, et al. Outcome after the modified Bentall technique with a long interposed graft to the left coronary artery. Ann Thorac Surg. 2009 Jan: 87(1):109-15.

39. Pomini F, et al. Cardiopulmonary bypass in pregnancy. Ann Thorac Surg 1996;61:259-68

40. Preiss M, Hösli I, Holzgreve W, Zerkowski HR Aortic dissection in pregnancy in Marfan syndrome--case report and treatment concept. Z Geburtshilfe Neonatol. 2001 May-Jun;205(3):110-3.

41. Robinson P N, Godfrey M. Marfan syndrome: a primer for clinicians and scientist. 2004: p 47

42. Rodeck C, Whittle M. Fetal medicine: basic science and clinical practice.2nd ed. 2009: p 143

43. Sakaguchi M, Kitahara H, Seto T, Furusawa T, Fukui D, Yanagiya N, Nishimura K, Amano J. Surgery for acute type A aortic dissection in pregnant patients with Marfan syndrome. *Eur J Cardiothorac Surg*. 2005 Aug ;28:280–283.

44. Sakaguchi M, Kitahara H, Watanabe T, Kono T, Fukui D, Amano J. Successful surgical treatment for acute aortic dissection in pregnancy with Marfan's syndrome. Jpn J Thorac Cardiovasc Surg. 2005 Apr;53(4):220-2.

45. Stecker MM, Cheung AT, Pochettino A, et al. Deep hypothermic circulatory arrest: I. Changes in electroencephalogram and evoked potentials. Ann Thorac Surg. 2001; 71:14-21

46. Stecker MM, Cheung AT, Pochettino A, et al. Deep hypothermic circulatory arrest: II. Changes in electroencephalogram and evoked potentials during rewarming. Ann Thorac Surg. 2001; 71:22-28

47. Svensson LG, Labib SB, Eisenhauer AC, et al. Intimal tear without hematoma: an important variant of aortic dissection that can elude current imaging techniques. Circulation. 1999;99:1331– 6.

48. Schwartz's principles of surgery, 8th edition, 2004.

49. Towsend. Sabiston Textbook of Surgery 18th edition, 2007

50. Tritapepe L, Voci P. Anesthesia for Caesarean section in a Marfan patient with recurrent aortic dissection. Can J Anesth 1996/43:11/pp 1153-5

51. Van Karnebeek CD, Naeff MS, Mulder BJ, et al. Natural history of cardiovascular manifestations in Marfan syndrome. Arch Dis Child.2001;84:129 –37.

52. White G. Basic Clinical Lab Competencies for Respiratory Care : An Integrated Approach. 2003. 4th ed : p 152

53. Winn H, Hobbins J. Clinical Maternal-Fetal Medicine. 2000 : 687

54. Zeebregts CJ, Schepens MA, Hameeteman TM, Morshuis WJ, de la Riviere AB. Acute aortic dissection complicating pregnancy. *Ann Thorac Surg.*1997;64:1345–134