Reviews

Aortic Dissection During Pregnancy: A Difficult Clinical Scenario

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ABSTRACT

Aortic dissection (AoD) during pregnancy is a rare but lethal condition and highlights the need for extensive elucidation. The aim of this study is to reveal the risk factors for AoD during pregnancy and to compare the 2 main risk factors, Marfan syndrome and pregnancy itself in the previously healthy woman. The pregnant patients developed AoD at 31.7 ± 7.6 weeks of gestation. It occurred much earlier in the Marfan patients than in the previously healthy women (30.7 ± 8.6 weeks of gestation vs 34.4 ± 4.4 weeks of gestation, $P = 0.0263$). In the Marfan patients, AoD developed in 3 (3.2%), 15 (15.8%), and 43 (46.3%) patients in the 3 trimesters, respectively, compared with 31 of the previously healthy women, and only in the third trimester. The neonates of the Marfan patients had better Apgar scores at 1 and 5 minutes, lower intubation rates, and fewer stays in the neonatal intensive care unit than those of the previously healthy women. Marfan syndrome and pregnancy itself in the previously healthy woman were the 2 main risk factors responsible for the occurrence of AoD during pregnancy. Marfan patients may develop AoD at an early age and an early stage of pregnancy, probably due to the preexisting weakened aortic wall. Better outcomes for the surviving neonates of Marfan patients compared with neonates of the previously healthy women might be the result of the poor condition of Marfan patients causing a higher death rate for those fetuses.

Introduction

Aortic dissection (AoD) during pregnancy is rare and life-threatening for both mother and fetus.[1] In Sweden it was estimated that the incidence of AoD was 14.5/1 000 000 in pregnant women vs 1.24/1 000 000 among nonpregnant women.[2] However, according to the International Registry of Acute Aortic Dissections, being in the peripartum period of pregnancy confers a risk for AoD of 0.2%.[3] Aortic dissection typically occurs in the third trimester of pregnancy or during the early postpartum period.[4] Aortic dissections often develop among individuals with connective-tissue disorders associated with abnormalities of the aortic wall, such as those present in familial thoracic aortic aneurysm/dissection, Marfan syndrome, Loeys-Dietz syndrome, vascular Ehlers-Danlos syndrome, and bicuspid aortic valve disease or Turner syndrome.[5] The majority of patients with Turner syndrome have aortic dilation with associated cardiovascular anomalies such as a bicuspid aortic valve, coarctation of the aorta, or systemic hypertension, which place the patients at higher risk of AoD. This is the case in particular when women with Turner syndrome, who are generally infertile because the condition is associated with premature ovarian failure, become pregnant through assisted reproductive technology.[6] Histologic findings are similar to the classic cystic medial necrosis among young patients with sporadic AoD and suggest an underlying genetic defect of connective tissue, resulting in dissection at an early age in such patients.[7] The risk of dissection is 5× to 18× higher in the presence of a bicuspid aortic valve compared with the tricuspid aortic valve, and pathology of the dilated aorta in the presence of a bicuspid aortic valve was similar to that of Marfan patients.[7] Parai et al[8] demonstrated significantly less elastic tissue in the aorta of bicuspid aortic valve patients. Women with aortic-root diameter >40 mm, rapid dilation of aortic dimensions, and/or previous dissection of the ascending aorta are at an increased risk of AoD during pregnancy.[9] In addition, the risks of complicated AoD may be increased in parturients in the presence of familial thoracic aortic aneurysm, bicuspid aortic valve, coarctation of the aorta, hypertension, and drug abuse.[10] However, the clinical characteristics of AoD during pregnancy have not been widely elaborated and the relationship between pregnancy and the risk factors for AoD remains insufficiently unexplained so far. This study aims to assess risk factors responsible for the development of acute AoD during pregnancy, comparing the 2 main underlying risk factors and discussing management controversies based on literature analyses.

Methods

Literature retrieval was made for English articles published from 1980 to 2012 concerning AoD and pregnancy using the MEDLINE database, Highwire Press, and Google search. The search terms were "aortic dissection" and "pregnancy."

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Articles describing AoD that developed during cesarean section or immediately after delivery were included. Articles reporting in general about pregnancy in relation to AoD with missing or obscure data were considered of no value for the statistical analysis and were thus omitted. Articles reporting aortic aneurysmal formation without dissection, or dissections of vessels other than the aorta during pregnancy, were excluded from this study. The literature search was terminated at 9:30 AM (Greenwich Mean Time + 8 hours) on January 1, 2013.

There are 2 classification schemes for AoD: DeBakey and Stanford. There are 3 types of AoD in the DeBakey classification: type I dissection, which originates in the ascending aorta and extends to at least the aortic arch; type II dissection, which involves the ascending aorta only; and type III dissection, which starts in the descending aorta, usually just distal to the left subclavian artery. The Stanford classification includes 2 types: type A dissection, which involves the ascending aorta; and type B dissection, which does not involve the ascending aorta.[5] The Stanford classification is used in the expression and analysis of AoD in this study.

Measurement data are reported as mean ± SD and compared with an unpaired *t* test. Comparisons of frequencies were made by Fisher exact test. A P value < 0.05 (2-tailed) was considered of statistical significance.

Results

The literature retrieval yielded 96 reports, comprising 85 case reports and 11 case series or original articles, with 122 patients.[5,11–105] Mean patient age (n = 120) was 31.6 ± 4.7 years (range, 19–43 years; median, 31.5 years) on admission. Patients were at 31.7 ± 7.6 weeks of gestation (range, 1–41 weeks; median, 34 weeks) at the onset of AoD. A history of pregnancy was reported in 78 patients: nulliparous in 42 (53.8%), primiparous in 19 (24.4%), and multiparous in 17 (21.9%). Of the latter, 8 (10.3%) were para 2; 3 (3.8%) were para 3; 1 each (1.3%, 1/80) were para 4, 5, and 9; and para number was not given in 3 (3.8%) patients.

The initial symptoms at the first onset of AoD were recorded in 92 patients. Pain was the most common symptom of AoD, which presented in 80 patients (87.0%): 79 (98.8%) had acute pain and 1 (1.2%) had mild pain. Chest pain was present in 56 patients (70%). Of those, the chest pains were characterized by retrosternal or parasternal locations in 5 patients (6.3%) and 2 patients (2.5%), respectively; radiating to the back in 4 patients (5%); and concurrent multisite pain including chest pain in 15 patients (16.3%). Back pain was the second onset symptom of AoD, which accounted for 21.7% (20/92) of all presentations. Epigastric and abdominal pain presented in 3 (3.8%) and 2 (2.5%) patients, respectively. Breast pain, neck pain with throat tightness, and leg with back pain were noted in 1 patient (1.3%, 1/80) each. One patient (1.1%, 1/92) had chest discomfort. Circulatory manifestations were seen in 9 (9.8%) patients: dyspnea in 4 (44.4%), persistent cough in 1 (11.1%), syncope in 1 (11.1%), and circulatory collapse in 3 (33.3%) patients, respectively. One patient (1.1%, 1/92) had neurological symptoms and 1 patient (1.1%, 1/92) was asymptomatic. Aortic dissection occurred during cesarean section in 1 patient[11] and immediately after delivery in 2 patients.[45,49]

In 4 patients (3.3%), AoD developed in the first trimester, at 5.7 ± 4.2 weeks of gestation (range, 1–9 weeks; median, 7 weeks). In 23 patients (18.9%), AoD developed in the second trimester, at 21.9 ± 3.8 weeks of gestation (range, 14–27 weeks; median, 22 weeks). Aortic dissection developed in the third trimester in 95 patients (77.9%), at 35.0 ± 3.6 weeks of gestation (range, 28–41 weeks; median, 36 weeks). Most of the patients were referred to hospital without delay after the acute onset and were admitted immediately. However, 15 patients had a delay from onset to admission of 360.9 ± 1295.6 hours (range, 2–5040 hours; median, 6 hours). Aortic dissection was immediately diagnosed at patient admission in all but 21 patients; 19 were diagnosed before labor and 2 after cesarean section.[13,44] In these patients, the diagnosis was delayed for 72.3 ± 105.7 hours (range, 5.5–408 hours; median, 10 hours). Surgical, interventional, or hybrid treatment was carried out in 99 patients. Time from the diagnosis of AoD to aortic operation was 54.6 ± 97.7 hours (range, 0.5–288 hours; median, 5.3 hours) in 14 patients (14.1%), whereas 85 patients (85.9%) received an urgent operation without delay.

Diagnostic modalities of AoD were reported in 90 patients. Computed tomographic scan and transthoracic and/or transesophageal echocardiography were the most common diagnostic tools, used in 32 (35.2%), 25 (27.5%), and 16 (17.6%) patients, respectively. Other diagnostic modalities included angiography in 6 (6.6%), magnetic resonance imaging in 4 (4.4%), and ultrasound (abdominal only) in 2 (2.2%) patients. The diagnosis was established by surgical exploration in 1 (1.1%, 1/92) patient and by autopsy in 5 (5.5%).

Types of AoD were reported in 118 patients: 92 patients (78.0%) had type A, 25 patients (21.2%) had type B (1 was associated with ascending aorta dilation), and 1 patient (0.8%) had simultaneous dissections of types A and B. The maximal diameter of the aorta was 54.1 ± 15.9 mm (range, 29–90 mm; median, 52 mm; n = 40). No significant difference was found in the maximal aortic diameter between type A and type B (56.2 ± 16.0 mm vs 46.3 ± 14.0 mm, $P = 0.1049$). Coronary ostia were involved by the AoD in 13 patients (10.7%); of these, 11 patients had involvement of 1 ostium and 2 patients had involvement of 2 ostia. A significant difference was noted in the prevalence between 1-ostium involvement and 2-ostia involvement (84.6% vs 15.4%, $P = 0.0012$). For cases involving 1 ostium, 7 were in the right, 1 was in the left, 1 was in noncoronary ostia, and in 2 cases the involved ostia were not mentioned. More right ostial involvements were found than involvements of the left or noncoronary ostia (77.8% vs 11.1% vs 11.1%, $P = 0.0024$). Annuloaortic ectasia was present in 6 patients (4.9%); of these, 3 patients (50%) had Marfan syndrome, 1 (16.7%) had Loeys-Dietz syndrome, and 2 (33.3%) had no underlying risk factors. The locations of the AoD entry tears were described in 18 patients: 17 patients (94.4%) had 1 entry tear, and 1 patient (5.6%) had 3 entry tears. The entry tears were located in the aortic root in 11 patients (61.1%): 5 (27.8%) above the aortic cusp, 1 (5.6%) above the coronary ostium, 3 (16.7%) in the sinus of Valsalva, and 2 (11.1%) in the sinotubular junction. Five entry tears (27.8%) were located in the ascending aorta. One entry tear (5.6%) was above the celiac artery. In 1 patient (5.6%), there were 3

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entry tears, located above the noncoronary cusp, above the right noncoronary commissure, and above the left aortic cusp, respectively.

Aortic-valve insufficiency was present as a result of AoD in 45 patients (36.9%): grade 1+ in 1 (2.2%), grade 2+ in 10 (22.2%), grade 3+ in 11 (49.4%), and grade 4+ in 23 (51.1%) patients, respectively. Mitral valve prolapse was present in 3 patients (2.5%). Pericardial and/or pleural effusions were present in 16 patients (13.1%), including 6 (37.5%) hemopericardia indicating a tamponade, 4 (25%) massive and 2 (12.5%) minor pericardium effusions, 1 (6.3%) pericardial and pleural effusion, and 3 (18.8%) pleural effusions.

The management of AoD was described in 115 patients. The majority of the patients underwent surgical operation, with stent-graft insertion, hybrid therapy, and conservative therapy in the minority. Eight patients who did not have prompt diagnosis or who presented with sudden death did not have an opportunity for surgical treatment (Table 1). During aortic operation, the arterial cannulation sites were recorded in 25 patients: the femoral artery in 20 patients (80%); axillary artery in 2 patients (8%); and brachiocephalic, carotid, and subclavian arteries in 1 patient (4%) each. The lowest temperature that was recorded in 36 patients was $22.8 \pm 5.9\,^{\circ}C$ (range, 11–34 °C; median, 22.1 °C). Six patients (16.7%) were at mild hypothermia of $32.5 \pm 0.8\,^{\circ}C$ (range, 32–34 °C; median, 32 °C); 7 patients (19.4%) were at moderate hypothermia of $26.7 \pm 1.6\,^{\circ}C$ (range, 25–28 °C; median, 28 °C); and 23 patients (63.9%) were at profound hypothermia of $19.1 \pm 3.2\,^{\circ}C$ (range, 11–24 °C; median, 19 °C). The cardiopulmonary bypass, cross-clamp, and circulatory arrest times were 209.9 ± 80.3 minutes (range, 74–372 minutes; median, 203 minutes; $n = 34$), 112.6 ± 48.3 minutes (range, 22–200 minutes; median, 109 minutes; $n = 26$), and 37.9 ± 22.8 minutes (range, 11–104 minutes; median, 31 minutes; $n = 21$), respectively. Cerebral perfusion was performed in 15 patients (12.3%) for the cerebral protection: 12 (80%) were antegrade and 3 (20%) were retrograde. Mothers and newborns were under a follow-up of 32.7 ± 38.4 months (range, 2–168 months; median, 17 months; $n = 35$) and 31.3 ± 40.4 months (range, 2–128 months; median, 12 months; $n = 11$), respectively.

Urgent or selective cesarean section was performed in 92 patients, 2 of whom were perimortem cesarean, at 34.6 ± 3.9 weeks of gestation (range, 24–41 weeks; median, 35 weeks). Vaginal birth was successful in 6 patients; 2 were instrumental vaginal deliveries. There were 8 occasions of fetal death (Table 2).

Aortic repair was performed at 30.7 ± 7.8 weeks of gestation (range, 1–40 weeks; median, 32 weeks; $n = 80$) for the pregnant women. Urgent cesarean section was carried out followed by aortic repair in a single session in 43 patients; aortic repair was performed at 2.2 ± 5.9 weeks (range, 4 hours–24 weeks; median, 0.35 weeks) after delivery in 17 patients; and urgent aortic repair with delayed selective cesarean section was performed in 16 patients, with an interval between aortic operation and cesarean section of 13.4 ± 9.0 weeks (range, 0.14–32 weeks; median, 15 weeks), respectively.

The body weight of the 53 newborns from 52 deliveries was 2452.4 ± 846.0 g (range, 950–4055 g; median, 2400 g).

Table 1. The Management of Aortic Dissection in 122 Patients

Management	No. (%)
Major aortic operations	93 (76.2)
Ascending aorta replacement	37 (40)
Bentall operation	29 (31.2)
Root replacement	12 (12.9)
Descending aorta replacement	5 (5.4)
David procedure	1 (1.1)
Aorta-bilateral iliac artery bypass	1 (1.1)
Surgical method not given	8 (8.6)
Stent-graft	4 (3.3)
Hybrid (ascending aorta stent, aortic valve resuspension)	1 (0.8)
Conservative treatment	8 (6.6)
Surgery for type A, conservative treatment for type B	1 (0.8)
Treatment method not given	7 (5.7)
No surgery due to sudden death or delayed diagnosis	8 (6.6)

Table 2. Modes of Delivery and Fetal Events in 108 Patients

Mode of Delivery and Fetal Event	No. (%)
Cesarean delivery	92 (85.2)
Usual cesarean	90 (83.3)[a]
Perimortem cesarean	2 (1.9)
Vaginal delivery	8 (7.4)
Spontaneous vaginal delivery	6 (5.6)
Instrumental vaginal delivery	2 (1.9)
Fetal event	8 (7.4)
Fetal abortion	1 (0.9)
Hysterotomy	1 (0.9)
Fetal demise	5 (4.6)
Fetal stillborn	1 (0.9)

[a] One pregnant patient delivered twin babies by cesarean section at 34 weeks of gestation.

Thirty-five newborns had their gender recorded; there were 23 males and 12 females, with a male-to-female ratio of 1.9:1. The Apgar scores at 1, 5, and 10 minutes were 4.9 ± 3.1 (range 0–10; median, 5; $n = 48$), 7.7 ± 2.0 (range, 2–10; median, 8; $n = 21$), and 6.5 ± 2.7 (range, 0–9; median, 7; $n = 12$), respectively. The umbilical pH was 6.99 ± 0.31 (range, 6.38–7.29; median, 7.00; $n = 8$).

There were 2 maternal morbidities, 1 due to progression of the AoD that required reoperation, and the other from subarachnoid hemorrhage. The mortalities of the mothers and fetuses were 16.4% (19/116) and 14.3% (17/112),

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Table 3. Maternal and Feto-Neonatal Outcomes in the 3 Trimesters

Outcome	First Trimester	Second Trimester	Third Trimester	*P* Value (Fisher Exact Test)
Maternal outcome, n (%)				
Survival	4 (100)	13 (76.5)	75 (83.3)	0.5585
Death	0 (0)	4 (23.5)	15 (16.7)	
Feto-neonatal outcome, n (%)				
Survival	3 (75)	14 (63.6)	79 (91.9)	0.0033
Death	1 (25)	8 (36.4)	7 (8.1)	

respectively. The neonate that required ventilation and had seizure activity was the only neonatal morbidity. The feto-neonatal other than maternal survival/mortality rates showed significant differences between different trimesters (Table 3).

Surgical specimens of the aorta were histologically studied in 29 patients. Medial myxoid changes were present in 15 (51.7%), cystic medial necrosis in 9 (31.0%), accumulation of acid mucopolysaccharides in 2 (6.9%), and collagenous disruption, elastic laminar disruption, and normal media in 1 (3.4%) each.

Marfan syndrome and pregnancy itself in the previously healthy woman were the 2 main risk factors leading to AoD in pregnancy (Table 4). There were no significant differences between the 2 subgroups in patient age at the time of AoD onset. Marfan patients developed AoD much earlier during pregnancy than did the previously healthy women. Marfan patients developed AoD in all 3 trimesters, most often in the third trimester, whereas the previously healthy women developed AoD only in the third trimester. There was an extremely significant difference in the frequencies of AoD in the 3 trimesters between the 2 subgroups ($P = 0.0006$) (Table 5).

A comparison of the maximal aortic diameter between <40 mm and ≥ 40 mm revealed a significant difference in the aortic diameter between the 2 subgroups (35.4 ± 2.9 mm vs 58.7 ± 14.1 mm, $P < 0.0001$). No intersubgroup differences were noted in other parameters, such as patient age, weeks of gestation at AoD onset, timing of cesarean section and aortic repair, type of AoD, incidence of AoD at different pregnancy stages, risk-factor distribution, and maternal and feto-neonatal outcomes.

Discussion

This study illustrated that hereditary fibrillinopathies represent the principal risk factors leading to AoD in pregnancy, with Marfan syndrome being the most common. One-quarter of the patients did not have any preexisting risk factors. Hereditary vascular diseases (bicuspid aortic valve and coarctation of the aorta), vascular inflammation disorders (Takayasu arteritis and aortitis), acquired heart disease (hypertension and coronary artery disease), and cocaine abuse can be associated with a considerable risk for the occurrence of AoD.

Table 4. Risk Factors Responsible for Aortic Dissection in 122 Parturients

Risk Factors	No. (%)
Hereditary fibrillinopathies	74 (60.7)
Marfan syndrome	61 (49.2)[a]
Turner syndrome	6 (4.9)
Familial thoracic aneurysm/dissection	4 (3.3)
Ehlers-Danlos syndrome	2 (1.6)
Loeys-Dietz syndrome	1 (0.8)
Previously healthy individual	31 (25.4)
Hereditary vascular diseases	7 (5.7)
Bicuspid aortic valve	6 (4.9)
Coarctation of the aorta	1 (0.8)
Vascular inflammation	3 (2.5)
Takayasu arteritis	2 (1.6)
Aortitis	1 (0.8)
Acquired heart disease	5 (4.1)
Hypertension	4 (3.3)
AMI	1 (0.8)
Others	2 (1.6)
Cocaine abuse	1 (0.8)
Antiphospholipid syndrome	1 (0.8)

Abbreviations: AMI, acute myocardial infarction.
[a] One patient had cocaine abuse.

It can be deduced that pregnancy alone with no underlying risk factors is an independent risk factor for AoD.[87] The hyperdynamic and hypervolemic characteristics of the parturients with tachycardia and enhanced stroke volume constitute the physiological basis of a predisposition to AoD.[39] Estrogen suppresses the synthesis of collagen and elastin.[106] Such hormonal effects may weaken the vascular walls, thereby playing an important role in the development of AoD.[22] Marfan, Turner, and Ehlers-Danlos syndromes are inherited connective-tissue disorders often characterized by disruption of the integrity of structural proteins, particularly of the aorta, affecting aortic composition and function so that patients are predisposed to aortic aneurysmal formation and dissection at early ages.[107]

A prospective study illustrated that women with Marfan syndrome with aortic dilation <40 mm tolerated pregnancy well, with good maternal and neonatal outcomes, whereas those with aortic-root diameters ≥ 40 mm at pregnancy seem to be at higher risk for aortic dilation, rupture, or dissection.[108] Aortic-root enlargement (≥ 40 mm) during pregnancy, the presence of a bicuspid aortic valve, and connective-tissue disorders are associated with considerable risk for type A AoD.[1] Familial thoracic aortic aneurysm has been identified with dominant inheritance, which may cause cystic medial necrosis in the absence of an

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Table 5. A Comparison Between Marfan Syndrome and Previously Healthy Women

Variables	Marfan Syndrome, n = 61	Previously Healthy Women, n = 31	*P* Value
Case no., n (%)	61 (50)	31 (25.4)	0.0001[a]
Age at the onset of AoD, y	31.3 ± 4.4	31.6 ± 4.9	0.7465[b]
AoD onset time, gestation wk	30.7 ± 8.6	34.4 ± 4.4	0.0263[a]
AoD during pregnancy, n (%)			
First trimester	3 (4.9)	0 (0)	0.0006[a]
Second trimester	15 (24.6)	0 (0)	
Third trimester	43 (70.5)	34 (100)	
Type of AoD, n (%)			
Type A	45 (73.8)	25 (86.2)	0.2782[a]
Type B	16 (26.2)	4 (13.8)	
Maximal dimension of the aorta, mm	55.1 ± 15.4	60.3 ± 26.3	0.6047[b]
AoD involvement, n (%)			
Coronary ostial involvement	8 (13.1)	3 (10.3)	0.7446[a]
Aortic valve regurgitation	22 (36.1)	17 (58.6)	0.0678[a]
Trivial	1 (4.5)	0 (0)	
Mild	4 (18.2)	5 (29.4)	0.1447[a]
Moderate	2 (9.1)	5 (29.4)	
Severe	15 (68.2)	7 (41.2)	
Annuloaortic ectasia	4 (6.6)	2 (6.9)	1.0000[a]
Mitral valve prolapse	3 (4.9)	0 (0)	0.5482[a]
Cardiopulmonary bypass for AoD repair			
Lowest core temperature, °C	23.5 ± 5.9	23.0 ± 6.3	0.8425[b]
Cardiopulmonary bypass time, min	232.4 ± 84.7	183.4 ± 75.9	0.1444[b]
Cross-clamp time, min	126.3 ± 42.4	94.3 ± 55.5	0.1646[b]
Circulatory arrest time, min	45.8 ± 28.4	28.2 ± 8.0	0.1623[b]
Modality of delivery (excluding fetal demise)			
Cesarean section, n (%)	42 (89.4)	22 (88)	
Postmortem cesarean section, n (%)	0 (0)	2 (8)	0.1189[a]
Vaginal delivery (including instrumental), n (%)	5 (10.6)	1 (4)	
Maternal mortality, n (%)	5 (8.2)	7 (22.6)	0.0972[a]
Fetus outcome			
Body weight, g	2538.4 ± 793.8	2517.6 ± 971.5	0.9413[b]
Apgar score at 1 min	5.7 ± 3.0	3.3 ± 2.7	0.0269[b]
Apgar score at 5 min	7.9 ± 1.9	6.0 ± 2.8	0.0250[b]
Intratracheal intubation and NICU stay, n (%)	5 (8.2)	9 (29.0)	0.0134[a]
Feto-neonatal death, n (%)	8 (13.1)	3 (9.7)	0.7446[a]

Abbreviations: AoD, aortic dissection; NICU, neonatal intensive care unit.
[a] Fisher exact test. [b] Unpaired *t* test. Data are presented as mean ± SD or n (%).

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associated syndrome[109] and is believed to be the cause of 20% of thoracic AoDs in general populations.[10] Clinical observations revealed that the dimensions of the aortic root and ascending aorta were larger in patients with a bicuspid aortic valve than in those with a tricuspid aortic valve with either normally functioning aortic valve,[110] or matched valvular lesions (aortic regurgitation, aortic stenosis, or mixed lesions),[111] or with aortic dilation.[112] Aortic-wall abnormalities were frequent in adults with coarctation of the aorta, although the ascending aorta in the patients with a bicuspid aortic valve and coarctation of the aorta did not dilate to the same degree as in the patients with isolated bicuspid aortic valve.[113] Immer et al[1] have compared types A and B AoDs in pregnant women. In a total of 45 women with acute type A dissection and 12 with type B dissection in pregnancy, type A dissection that developed prepartum extended 48 mm in diameter, whereas the aortic dimensions of type B dissection were not measured. Compared with Marfan patients, the pregnant patients with a bicuspid aortic valve were younger and dissection developed in an earlier gestation. However, the authors did not include previously healthy women in the evaluation. Of special interest, 1 patient included in this study had 2 segmental AoDs in the ascending aorta and descending aorta simultaneously, which was taken as a type A + B AoD by the authors.[103] However, it might be categorized as type A. Takayasu arteritis most commonly involves the aortic arch and its major branches, leading to aortic-wall weakening and eventual aneurysmal formation or dissection.[114] Cocaine seems to predispose patients to acute AoD. With cocaine abuse, blood pressure surges and forces the blood between the intima and outer layers of the aorta, allowing a cocaine-induced intimal injury to precipitate the AoD process.[115]

It has been reported that pain was the common symptom or complaint at the onset of AoD, with back pain accounting for 55% of all presentations and chest pain accounting for 12%.[116] The present patient setting manifested pain in 87% of all presentations, with chest pain representing 70% (60.9% of all presentations), and back pain, 20% (17.4% of all presentations). In almost half of the chest-pain patients, the chest pain was characterized by retrosternal or parasternal locations, or radiating to the back, or multisite pain. The pain can be sharp, tearing, or ripping. The patients' symptoms may arouse the consulting physicians' suspicion of AoD. Echocardiography, computed tomography, and magnetic resonance imaging are important noninvasive means of ensuring the diagnosis of acute AoD, necessitating a differential diagnosis from acute pulmonary embolism. Intimal flaps, true and false lumens, and entry tears are typical signs of AoD, whereas echocardiographic visualization of pericardial effusions and intramural hematomas can be indirect signs pointing to the diagnosis of AoD.[92,117,118] Approximately 65% of intimal tears occur in the ascending aorta, 30% occur in the descending aorta, less than 10% occur in the aortic arch, and approximately 1% occur in the abdominal aorta.[119] Adequate investigation, prompt diagnosis, and timely management of AoD are crucial to reduce the mortality and morbidity of both mother and fetus. Delay of intervention with a type A AoD is directly correlated with considerable mortality, with an increment of 1% per hour.[116]

By comparing the 2 main risk factors, Marfan syndrome and pregnancy alone in the previously healthy women, one can find that, during pregnancy, women with Marfan syndrome may develop AoD in all 3 trimesters, most often in the third trimester, whereas in the previously healthy women, AoD developed only in the third trimester. The onset of AoD in Marfan syndrome parturients was 3.5 weeks earlier than that in the previously healthy pregnant women. Similar to the previous report, Marfan patients developed AoD 7.7 years earlier than non-Marfan patients.[120] This can be explained by the more significant hemodynamic instability due to inherent collagen deficiency in Marfan patients than in normal human subjects.[120] The newborns from the mothers with Marfan syndrome had higher Apgar scores than those from previously healthy mothers and had fewer requirements for intratracheal intubation and stays in the neonatal intensive care unit. It seemed that the higher number of cesarean sections performed on the previously healthy women compared with Marfan patients would lead to better fetal outcomes, as scheduled cesarean section allows for better control of hemodynamic parameters.[121] However, the fact could not be overlooked that Marfan patients had a much higher fetal-demise rate during very early periods of pregnancy. Therefore, the surviving fetuses of the Marfan patients showed better postpartum results.

The treatment depends on the site of the AoD, with emergency surgery recommended for acute type A dissections and conservative therapy for type B dissections.[119] Upon diagnosis of AoD, immediate intravenous nitroprusside and a β-blocker should be initiated,[38] and surgical intervention is mandatory for type A AoD. When aortic-root dimension is <40 mm, vaginal delivery can be tolerated, whereas cesarean section is warranted in women whose aortic-root dimension is >40 mm.[3] Therefore, women with aortic-root dimension >40 mm should avoid pregnancy.[122] The timing of aortic repair and delivery have to be determined based on the stage of pregnancy and the status of the fetus. Aortic repair with continued pregnancy is recommended for the parturient before 28 weeks of gestation; between 28 and 32 weeks of gestation, pregnancy should be prolonged if the aortic repair is tolerated; after 32 weeks of gestation, primary cesarean section followed by aortic repair performed in a single operative session is recommended; and immediate delivery is mandatory in the event of fetal distress.[35] However, fetal survival in the setting of cardiopulmonary bypass or profound hypothermic circulatory arrest cannot be anticipated in advance.[123] The mother's health should prevail over fetal concerns.[37]

Aortic dissection in pregnancy is a complex clinical scenario, difficult to diagnose and difficult to treat, considering the survival of both mother and fetus. The present study revealed urgent cesarean section followed by aortic repair in a single session was needed in 65.3% of cases, urgent aortic repair with delayed selective cesarean section in 16.3% of cases, and aortic repair was performed 3 weeks after the delivery in 18.4% of cases. The reason for urgent cesarean section for the pregnant woman with AoD was to avoid hemodynamic stress, progressive aortic expansion, and pending aortic rupture.[124] Elsewhere, urgent cesarean section may

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also be due to fetal distress,[39,44,78] fetal intrauterine growth retardation,[71] and abnormal presentations.[101]

The missing data of some patients constitute the main drawback of this study, making for less significant differences between subgroups. Abundant information collection may facilitate precise statistical analysis in the long run.

Conclusion

The most common underlying cause responsible for the development of AoD in the pregnant woman was Marfan syndrome, followed by pregnancy itself in the previously healthy pregnant woman without any other predisposing risk factors. Marfan patients may develop AoD at an early age and an early stage of pregnancy, probably due to the preexisting weakened aortic wall. Better outcomes for the surviving neonates of Marfan patients compared with neonates of the previously healthy women might be the result of the poor condition of Marfan patients causing a higher death rate for those fetuses. Prompt diagnosis and subsequent proper management may save the lives of both the mother and fetus.

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