Clinical Report

Treatment of patients with aortic disease during pregnancy and after delivery

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Abstract
Objective: This study aimed to review treatment of patients with aortic disease during pregnancy and after delivery in Beijing Anzhen Hospital.

Methods: A retrospective study was conducted using data from 12 patients with aortic disease during pregnancy and after delivery in our institution from May 2005 to December 2014. Patients were provided different treatments based on the type of aortic disease and clinical characteristics.

Results: The mean age was 29.83 ± 4.17 years, mean height was 171.7 ± 8.22 cm, mean weight was 68.55 ± 10.62 kg, and mean body mass index was 23.18 ± 2.93 kg/m². Two patients with Stanford type A aortic dissection died of renal failure after surgery. All of the other patients were discharged. Six foetuses survived. One patient continued her pregnancy after an aortic operation. The gestational age of the remaining five patients was less than 28 weeks at the time of the operation and all foetuses of these five patients died.

Conclusions: A suitable treatment strategy for aortic disease during pregnancy and after delivery should be chosen based on an individual’s comprehensive clinical condition. Foetal management should be chosen based on gestational age and severity of aortic disease.

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Keywords
Pregnant patient, aortic disease, pregnancy, delivery, treatment

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Introduction
Aortic disease during pregnancy and after delivery is an extremely rare group of serious cardiovascular diseases that include aortic dissection and aortic aneurysm.1 Aortic dissection and aortic aneurysm can lead to a significantly higher risk of maternal mortality in pregnant women and are often associated with catastrophic outcomes.2 If these conditions occur before delivery, the foetus might die.1 Approximately half of all female patients younger than 40 years old with aortic dissection present with this disease in the perinatal period. Aortic disease usually occurs in the third trimester or early post-partum period. After reviewing the published literature, 96 articles on the diagnosis and treatment of aortic dissection during pregnancy and after delivery were searched. These articles included 85 case reports and 11 case series that reported 122 patients with aortic dissection during pregnancy and after delivery. The types of aortic dissection were reported in 118 of these patients; 93 (78.8%) patients had Stanford type A and 25 (21.2%) had Stanford type B.2 Marfan syndrome plays an important role in aortic dissection in pregnancy. Endovascular treatment should not be used in patients with Marfan syndrome. Four patients with aortic root aneurysm were reported in pregnancy.1 Approximately 50% of aortic dissections in young female patients are associated with pregnancy.3 When treating patients with aortic disease during pregnancy, cardiac surgeons have to simultaneously consider rescuing the patient and her foetus.1 We performed a retrospective study using data from patients with aortic disease during pregnancy and after delivery to assess different methods of treating these patients in our institute. The results of this study will hopefully play a guiding role in the future treatment of such patients.

Methods
The institutional review board of Beijing Anzhen Hospital approved the design of this study. Informed consent of all patients was obtained for this study. Twelve pregnant patients with aortic disease during pregnancy and after delivery (the longest post-partum period was 6.5 months) at our institution from May 2005 to December 2014 (Table 1, patient nos. 1–12) were included in the study. All of the patients received preoperative computed tomographic angiography (CTA) and transthoracic echocardiography, which are the two most important diagnostic tests for pregnant patients with aortic diseases.2 However, some of the diagnoses of Stanford type A aortic dissection by transthoracic echocardiography were proven as false during surgery. Therefore, even if there was the possibility of harming the foetus, we still insisted that CTA should be performed in all of the patients.

According to the Stanford aortic dissection classification,4 preoperative CTA and echocardiography results showed that five patients had Stanford type A aortic dissection with aortic regurgitation and pathology of the aortic arch (three of them had acute dissection, Figure 1(a)). Two of these patients suffered from Marfan syndrome. We diagnosed Marfan syndrome in this study according to a clinical examination, as described in previous studies5,6). Three of these patients suffered from hypertension. The diagnosis of hypertension in this study
Table 1. Basic clinical information before surgery.

| Patient number | Age (y) | Height (cm) | Weight (kg) | BMI (kg/m²) | Aortic sinus diameter (mm) | Ascending aortic diameter (mm) |
|----------------|---------|-------------|-------------|-------------|---------------------------|-------------------------------|
| 1              | 27      | 175         | 76          | 24.81633    | 81                        | 48                            |
| 2              | 27      | 178         | 78          | 24.6181     | 61                        | 60                            |
| 3              | 32      | 176         | 70          | 22.5981     | 62                        | 63                            |
| 4              | 35      | 167         | 75          | 26.89232    | 42                        | 41                            |
| 5              | 34      | 172         | 80          | 27          | 42                        | 49                            |
| 6              | 27      | /           | /           | /           | 33                        | 35                            |
| 7              | 22      | 168         | 70          | 24.8016     | 29                        | 24                            |
| 8              | 35      | 160         | 60          | 23.4        | 35                        | 33                            |
| 9              | 25      | 173         | 55          | 18.4        | 42                        | 26                            |
| 10             | 31      | 175         | 69          | 22.53       | 70                        | 66                            |
| 11             | 31      | 158         | 46          | 18.43       | 38                        | 57                            |
| 12             | 32      | 187         | 75          | 21.44757    | 45                        | 32                            |

Figure 1. (a) Preoperative CTA and (b) postoperative CTA results showing Stanford type A aortic dissection in a pregnant patient.
was consistent with the “Diagnosis and treatment guideline of hypertensive disorders in pregnancy (2015)”, which was published by Hypertensive Disorders in Pregnancy Subgroup, Chinese Society of Obstetrics and Gynecology, Chinese Medical Association7. Additionally, one of these patients had dissection of the opening of the right coronary artery.

Two patients had a simple Stanford type B aortic dissection (both of them had hypertension and non-Marfan syndrome). Two patients had a complicated Stanford type B aortic dissection, whose left subclavian arteries were affected by the dissection, but without end-organ ischaemia.8 One of these two patients had Marfan syndrome and had dissection and dilation of the total thoracic and abdominal aorta. The other patient with hypertension suffered from a pathology around the left subclavian arteries. Two patients had a simple aortic root aneurysm, and one patient with Marfan syndrome had an aortic root aneurysm combined with Stanford type B aortic dissection (Tables 1, 2). Because of the patients’ age, we implanted mechanical aortic valve prostheses in all of those who underwent the Bentall procedure. Each foetus was delivered via caesarean section or abortion before, after, or intra-operation.

Results

The mean age of the included patients was 29.8 ± 4.2 years old. The mean height was 171.7 ± 8.2 cm. The mean weight was 68.6 ± 10.6 kg and the mean body mass index (BMI) was 23.2 ± 2.9 kg/m².

1. Type A aortic dissection during pregnancy and after delivery

All five patients with type A aortic dissection in this study received the Bentall procedure and total arch replacement using a tetrafurcate graft and stented elephant trunk implantation9 (Figure 1(b)). Taking the patient’s pregnant condition into consideration, we needed to reduce the time of cardiopulmonary bypass as much as possible. Therefore, we decided to perform the Bentall procedure and total arch replacement in these five patients to avoid unnecessary surgery time from a possible aortic valve replacement after unsatisfactory aortic valvuloplasty. We performed a right coronary artery bypass in a patient who had dissection of the opening of the right coronary artery. All five patients received antegrade cerebral perfusion through the right axillary artery.

Three of these five patients had acute aortic dissection, and received surgery within 24 h after presentation. The other two patients had chronic aortic dissection. One of these two patients received surgery 40 days after presentation and the other patient received surgery 24 h after presentation, but both intraoperative findings showed chronic aortic dissection. One patient at 22 + weeks of gestation and one at 34 + weeks had caesarean section and hysterectomy. One patient at 29 + weeks of gestation and one at 32 + weeks had caesarean section without hysterectomy. One patient underwent aortic surgery after caesarean section. The mean time of cardiopulmonary bypass (CPB) was 183.40 ± 15.40 min. The mean time of aortic clamping was 96.18 ± 13.54 min and the mean time of the deep hypothermic circulatory arrest was 30.60 ± 11.70 min (Table 2, patient nos. 1–5). Two patients suffered from renal failure and other complications after the operation. Both of these patients died from multiorgan failure associated with aortic surgery. One patient had caesarean section and hysterectomy at 22 + weeks of gestation at the same time as the aortic operation, and the foetus died. One patient received aortic surgery 2 months after caesarean section, and the newborn was well and scored 10-10-10
| Number | Aortic condition | Gestation at presentation (weeks) | Background condition | Timing of aortic surgery | Type of aortic surgery | Patient outcome | Obstetric management | Foetal outcome |
|--------|-----------------|----------------------------------|----------------------|--------------------------|------------------------|-----------------|---------------------|---------------|
| 1      | Stanford type A aortic dissection with aortic regurgitation and pathology of the aortic arch | 29 | During pregnancy | Bentall + total arch replacement by a tetrafurcate graft and stented elephant trunk implantation | Recovered | Caesarean section | Alive, Apgar scores of 4 (1 min), 4 (5 min), and 6 (10 min) |
| 2      | Stanford type A aortic dissection with aortic regurgitation and pathology of the aortic arch | 32 | Marfan | During pregnancy | Bentall + total arch replacement by a tetrafurcate graft and stented elephant trunk implantation | Recovered | Caesarean section | Alive, Apgar scores of 5 (1 min), 8 (5 min), and 9 (10 min) |
| 3      | Acute Stanford type A aortic dissection with aortic regurgitation and pathology of the aortic arch | 22 | Marfan, hypertension | During pregnancy | Bentall + total arch replacement by a tetrafurcate graft and stented elephant trunk implantation + CABG | Deceased | Caesarean abortion and hysterectomy | Stillbirth |
| 4      | Acute Stanford type A aortic dissection with aortic regurgitation and pathology of the aortic arch | 34 | Hypertension | During pregnancy | Bentall + total arch replacement by a tetrafurcate graft and stented elephant trunk implantation | Recovered | Caesarean section and hysterectomy | Alive, Apgar scores of 4 (1 min), 5 (5 min), and 9 (10 min) |
| 5      | Acute Stanford type A aortic dissection with aortic regurgitation and pathology of the aortic arch | 38 | Hypertension | Post-partum | Bentall + total arch replacement by a tetrafurcate graft and stented elephant trunk implantation | Deceased | Caesarean section 2 months before the operation | Alive, Apgar scores of 10 (1 min), 10 (5 min), and 10 (10 min) |
| 6      | Simple Stanford type B aortic dissection | 38 | Hypertension | Post-partum | Conservative treatment | Recovered | Caesarean section 40 days before admission to hospital | Alive, Apgar scores of 10 (1 min), 10 (5 min), and 10 (10 min) |
| 7      | Acute simple Stanford type B aortic dissection | 28 | Hypertension | During pregnancy | Aortic endovascular repair | Recovered | Caesarean section 2 weeks after the operation | Alive, Apgar scores of 10 (1 min), 10 (5 min), and 10 (10 min) |
| 8      | Complicated Stanford type B aortic dissection | 23 | Hypertension | Post-partum | Left common carotid artery to left subclavian artery bypass + stented elephant trunk implantation | Recovered | Water ball to induce labour 27 days before the operation | Stillbirth |
One patient at 34+ weeks of gestation who received caesarean section and hysterectomy at the same time as the aortic operation, and two patients at 29+ weeks and 32+ weeks of gestation who received caesarean section without hysterectomy, recovered well without surgery-related complications. Their newborns had Apgar scores of 4-4-6, 5-8-9, and 4-5-9 (1 min-5 min-10 min), respectively, and all were discharged uneventfully after treatment (Table 2, patient nos. 1–5).

2. Type B aortic dissection during pregnancy and after delivery

We treated two patients with simple type B aortic dissection during pregnancy. Patients with simple type B aortic dissection should receive aortic endovascular repair (EVAR).3,10 One patient in this study who was 28+ weeks' gestation received EVAR. However, the other patient only received conservative treatment 40 days after caesarean section because her femoral artery was too thin to receive EVAR (Table 2, patient nos. 6, 7). The patient who received EVAR recovered uneventfully. Two weeks after EVAR, she gave birth and her foetus was well and had an Apgar score of 10-10-10 (1 min-5 min-10 min). The other patient's condition improved after a period of conservative treatment. She had an Apgar score of 8-10-10 (1 min-5 min-10 min) 27 days before the aortic operation and was discharged after a period of conservative treatment. Her foetus was well and had an Apgar score of 10-10-10 (1 min-5 min-10 min) 27 days before the aortic operation. She had a caesarean section 40 days before visiting the hospital, and her foetus was well and had an Apgar score of 10-10-10 (1 min-5 min-10 min).

One patient (patient no. 8) with pathology of the aortic arch had a gestational age of 23+ weeks. Twenty-seven days after using a water ball to induce labour, she gave birth and her foetus was well and had an Apgar score of 3.5 months after the operation. One patient at 34+ weeks of gestation who received caesarean section and hysterectomy at the same time as the aortic operation, and two patients at 29+ weeks of gestation who received caesarean section without hysterectomy, recovered well without surgery-related complications. Their newborns had Apgar scores of 4-4-6, 5-8-9, and 4-5-9 (1 min-5 min-10 min), respectively, and all were discharged uneventfully after treatment (Table 2, patient nos. 1–5).

Table 2. Continued.

| Number | Aortic condition | Gestation at presentation (weeks) | Background condition | Timing of aortic surgery | Type of aortic surgery | Patient outcome | Obstetric management | Foetal outcome |
|--------|------------------|----------------------------------|----------------------|--------------------------|------------------------|-----------------|----------------------|---------------|
| 9      | Acute complicated Stanford type B aortic dissection | 22 | Marfan | Post-partum | Total thoracoabdominal aortic replacement | Recovered | Caesarean abortion and double tubal resection 3.5 months before the operation | Stillbirth |
| 10     | Aortic root aneurysm | 18 | Hypertension | During pregnancy | Bentall procedure | Recovered | Continued pregnancy after the Bentall procedure | Continued pregnancy |
| 11     | Aortic root aneurysm | 27 | Post-partum | Bentall procedure | Recovered | Caesarean abortion 6.5 months before the aortic operation | Stillbirth |
| 12     | Aortic root aneurysm combined with Stanford type B aortic dissection | 8 | Marfan, hypertension | Post-partum | Bentall + total arch replacement by a tetrafurcate graft and stented elephant trunk implantation | Recovered | Vacuum aspiration 27 days before the aortic surgery | Stillbirth |
subclavian artery bypass and stented elephant trunk implantation. Antegrade cerebral perfusion through the right axillary artery was used. The other patient (patient no. 9) had a gestational age of 22 + weeks and had dissection and dilation of the total thoracoabdominal aorta. Because of Marfan syndrome, she received a total thoracoabdominal aortic replacement. Three and a half months after caesarean section and double tubal resection, she underwent this kind of treatment. Patient no. 8 had pathology of the aortic arch and had received a water ball to induce labour 27 days before she visited hospital, and was 23 + weeks of gestation. During aortic surgery, the times of CPB, aortic clamping, and deep hypothermic circulatory arrest were 86 min, 37 min, and 17 min, respectively (Table 2). After the aortic surgery, she was discharged with no significant complications. The other patient suffered from dissection and dilation of the total thoracoabdominal aorta and was 22 weeks of gestation. During surgery, the femoral artery and vein were used to perform CPB. The duration of CPB was 112 min (Table 2, patient no. 9). She received caesarean section and double tubal resection 3.5 months before the total thoracoabdominal aortic replacement. After the aortic operation, she recovered uneventfully. Both of the foetuses died.

3. Simple aortic root aneurysm after delivery

We treated two patients with a simple aortic root aneurysm during pregnancy and after delivery. Based on the pathological anatomy of aortic root aneurysms, the convention is to replace the aortic valve and aortic root, and transplant the left and right coronary arteries (Bentall procedure). Therefore, we used the Bentall procedure for these two patients. The mean times of CPB and aortic clamping were 100 ± 26.9 min and 73.5 ± 30.4 min, respectively. One patient had a gestational age of 18 + weeks. Her foetus was normal and she continued pregnancy after the Bentall procedure. The other patient had a gestational age of 27 + weeks. Her foetus was delivered via caesarean section 6.5 months before the aortic operation, but her foetus died (Table 2, patient nos. 10, 11). Both patients recovered without surgery-related complications, and the results of the aortic pathology were both myxoid changes (Table 2, patient nos. 10, 11).

4. Aortic root aneurysm combined with Stanford type B aortic dissection during pregnancy and after delivery

We treated one patient with an aortic aneurysm combined with Stanford type B aortic dissection during pregnancy and after delivery. This patient’s aortic aneurysm involved the aortic root and arch. The tear site was located near the left subclavian artery. Therefore, we decided to perform the Bentall procedure and total arch replacement using a tetrafurcate graft and stented elephant trunk implantation. The times of CPB, aortic clamping, and deep hypothermic circulatory arrest were 224 min, 172 min, and 28 min, respectively. This patient had a gestational age of 8 + weeks. She had vacuum aspiration 27 days before aortic surgery (Table 2, patient no. 12). Her foetus died. However, she recovered uneventfully after the Bentall procedure and total arch replacement (Table 2, patient no. 12).

Discussion

Aortic disease during pregnancy and after delivery can significantly increase the risk of foetal and maternal mortality. Stanford type A and Stanford type B aortic dissections are involved in perinatal aortic dissection. In women younger than 40 years old, 50% of aortic dissections and aortic
Aneurysms are associated with pregnancy.\textsuperscript{1,3} They usually occur in the third trimester of pregnancy.\textsuperscript{1,3} In the present study, approximately half of the patients suffered from aortic disorders in the third trimester of pregnancy, which suggested that more attention should be paid to pregnant women in this period. Age is another risk factor of aortic disease. All of the patients in this study were younger than 35 years old, which is consistent with the conclusion of previous studies that young pregnant women are more prone to aortic disease.\textsuperscript{1,3} Other risk factors responsible for aortic disease include a history of hypertension and Marfan syndrome.\textsuperscript{2,11} A total of 49.5\% patients with aortic dissection during pregnancy and after delivery have Marfan syndrome, and 3.3\% of patients have hypertension.\textsuperscript{2} In our study, there were only two patients without both hypertension and Marfan syndrome, which is a similar finding to previous studies.

The total mortality of mothers in the present study was 16.7\%. All of the foetuses died in patients with Stanford type A aortic dissection. In fact, death caused by Stanford type A aortic dissection accounts for 14\% of cardiac deaths in pregnant women.\textsuperscript{12} Approximately 87.5\% of Stanford type A aortic dissections occur before delivery and usually have catastrophic outcomes.\textsuperscript{1} The mortality of acute Stanford type A aortic dissection in this study was over 66\%. This finding suggested that acute Stanford type A aortic dissection was the most severe aortic disorder during pregnancy and after delivery. All patients with chronic Stanford type A aortic dissection survived in this study. This result is consistent with the conclusions of previous studies that surgery on patients with chronic type A dissection is safer than that in those with acute type A dissection.\textsuperscript{13,14}

All patients with type A aortic dissection have lesions of the aortic valve and arch.\textsuperscript{4} These patients should receive the Bentall procedure, valve-sparing and ascending aortic replacement, or only ascending aortic replacement for different aortic valve disorders.\textsuperscript{3} All patients with type A aortic dissection in this study had severe aortic regurgitation. Therefore, all of these patients received the Bentall procedure and total arch replacement using a tetrafurcate graft and stented elephant trunk implantation.

Stanford type B aortic dissection affects approximately 60\% of patients before delivery and approximately 40\% in the early period after delivery.\textsuperscript{2} Some of these patients also have an aortic root aneurysm.\textsuperscript{1} Strategies for this condition include conservative treatment, surgery, and EVAR \textsuperscript{13}. Previous studies have reported that mortality of conservative and surgical treatment for patients with Stanford type B aortic dissection during pregnancy and after delivery ranges from 30.8\% to 42.8\%.\textsuperscript{1,15} EVAR treatment is widely used in pregnancy-related Stanford type B aortic dissections,\textsuperscript{10,16} which yields satisfactory results in pregnancy.\textsuperscript{15} For patients with Stanford type B aortic dissection who are qualified for EVAR during pregnancy and after delivery, we first recommend endovascular treatment for avoiding surgical injury to pregnant women. For patients with complications or other contraindications, we need to perform surgical or conservative treatment. EVAR should not be used in patients with Marfan syndrome. Therefore, patients with acute type B aortic dissection and Marfan syndrome receive total thoracoabdominal aortic replacement.

Aortic root aneurysm usually occurs in the third trimester of pregnancy.\textsuperscript{5} For these patients, if aortic lesions can be tolerated, we recommend conservative treatment. However, the two patients with an aortic root aneurysm who were included in the present study were extremely critical. Based on the experience of these patients in the present study, treatment for these patients is similar to that for Stanford type A aortic dissection, and requires surgery.
In this study, six foetuses survived. One patient continued her pregnancy after the aortic operation. The gestational ages of the remaining five patients were less than 28 weeks. Therefore, management of foetuses should be decided based on gestational age. For patients with a pregnancy of less than 28 weeks, they should begin treatment for aortic disease after an abortion. However, if their condition is critical, such patients with Stanford type A aortic dissection should receive aortic surgery first and then decide to continue or stop the pregnancy. For patients with more than a 28-week pregnancy, an attempt should be made to save the foetus. If the aortic condition is stable, patients should start treatment after caesarean section. If this is not the case, the patient should receive aortic surgery and caesarean section simultaneously.

In summary, determining an optimal treatment for aortic disease during pregnancy and after delivery requires comprehensive judgment based on various clinical conditions and therapeutic indications. Patients with acute Stanford type A aortic dissection have the worst prognosis. We recommend that surgical intervention should be delayed as much as possible in pregnant women with acute Stanford type A aortic dissection. For patients with Stanford type B aortic dissection who qualify for EVAR, an endovascular treatment is recommended. Moderate aortic aneurysm is recommended to receive conservative treatment. However, treatment for a critical ascending aortic aneurysm is similar to that for Stanford type A aortic dissection and requires surgery. Finally, foetal management should be chosen based on gestational age and severity of the aortic disease.

Declaration of conflicting interest
The Authors declare that there is no conflict of interest.

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