Aortic Dissection in Pregnancy: Management Strategy and Outcomes

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Background. **Aortic dissection in pregnancy is a rare but lethal catastrophe. Clinical experiences are limited. We report our experience in 25 patients focusing on etiology, management strategies, and outcomes.**

Methods. **Between June 1998 and February 2015, we treated 25 pregnant women (mean age, 31.6 ± 4.7 years) in whom aortic dissection developed at a mean of 28 ± 10 gestational weeks (GWs). Type A aortic dissection (TAAD) was present in 20 (80%) and type B (TBAD) in 5 (20%). Marfan syndrome was seen in 17 (68%). Management strategy was based on dissection type and GWs.**

Results. **TAADs were managed surgically in 19 (95.0%) and medically in 1 (5.0%). Maternal and fetal mortalities were, respectively, 14.3% (1 of 7) and 0 (0 of 7) in the "delivery first" group (7 of 20), 16.7% (1 of 6) and 33.3% (2 of 6) in "single-stage delivery and aortic repair" group (6 of 20), 16.7% (1 of 6) and 66.7% (4 of 6) in "aortic repair first" group (6 of 20), and 100% (1 of 1) and 100% (1 of 1) in the "medical management" group (1 of 20). TBADs were managed surgically in 60% (3 of 5) and endovascularly and medically in 20% each (1 of 5). No maternal deaths occurred. Fetal mortality was 100% in the surgical group and 0% in the other groups. During late follow-up, which was complete in 95.2% (20 of 21), 3 maternal and 2 fetal deaths occurred in the TAAD group. Overall maternal survival was 68.6% at 5 years.**

Conclusions. **Marfan syndrome predominates among women with aortic dissection in pregnancy. For TAADs, after 28 GWs, delivery followed by surgical repair can achieve maternal and fetal survival adequately; before 28 GWs, maternal survival should be prioritized given the high risk of fetal death. For TBADs in pregnancy, nonsurgical management is preferred.**

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Aortic dissection associated with pregnancy is a particularly unique clinical catastrophe that is potentially devastating to the mother and her fetus. Aortic dissection is partly attributable to the physiologic effects of pregnancy [1] and may occur in the 3 trimesters and postpartum period. Reports from the International Registry of Acute Aortic Dissection [2] and other population-based studies [3–5] have shown that aortic dissection in pregnancy is an extremely rare occurrence, accounting for 0.1% to 0.4% of all aortic dissections [2, 4] and representing 0.0004% of all pregnancies between 1998 and 2008 in the Nationwide Inpatient Sample database [4].

To date, clinical experience with this entity is limited to case reports [6–11] or small cohorts containing fewer than 20 patients [12–16]. A search of the English literature revealed approximately 180 cases since 1944 [6, 14, 17]. This report describes our experiences in management of 25 pregnant women with aortic dissection, focusing on etiology, treatment strategies, and outcomes, to aid cardiac surgeons in the management of this rare but challenging problem.

Patients and Methods

The Ethics Committees of Beijing Anzhen Hospital of Capital Medical University approved this retrospective study.

Patients

Between June 1998 and February 2015, our group treated 25 women with aortic dissection associated with pregnancy. Among these, 15 patients were managed in our

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own institutions and 10 patients in 9 other hospitals across China (as listed in the Acknowledgment). Mean age was 31.6 ± 4.7 years (median, 31; range, 24 to 44 years). The mean gestational age at aortic dissection was 28 ± 10 gestational weeks (GWs; range, 6 GWs to 6 weeks postpartum). Aortic dissection occurred in the first, second, and third trimesters and in the postpartum period in 8.0% (2 of 25), 36% (9 of 25), 36% (9 of 25), and 20% (5 of 25), respectively (Table 1 and Supplementary Table 1). A type A aortic dissection (TAAD) occurred in 20 patients (80.0%; Figs 1 and 2) and type B (TBAD) in 5 (20%).

All diagnoses were made by computed tomographic (CT) angiography. Marfan syndrome (by Ghent criteria) was seen in 17 patients (68.0%). Hypertension was present in 7 patients (28.0%), 3 of whom were diagnosed with Marfan syndrome and 4 without. Moderate to severe aortic regurgitation was present in 19 patients (76%), predominantly in the TAAD group (18 of 19). The diameter of the aortic root was 54 ± 15 mm.

Management Strategy

Surgical, endovascular, or medical management was determined by the type of aortic dissection and gestational age (delivery first or aortic repair first). In the 20 patients with TAAD, management was surgical in 19 (95%) and medical in 1 (15%). In the 5 patients with TBAD, management was surgical in 3 (60%) and medical and endovascular in 1 (20%) each.

The sequences of aortic repair and delivery were: delivery, followed by aortic repair in two stages, in 10 patients (40.0%); single-stage delivery, followed by aortic repair, in 6 (24.0%); and aortic repair, followed by delivery, in 7 (28.0%). After cesarean delivery, a Cook balloon was inserted in the uterus to prevent postoperative bleeding (Table 2).

Table 1. Patients' Profiles

Variables[a]	Aortic Dissection		
	Type A	Type B	Total
Patients	20 (80)	5 (20)	25 (100)
Age, y	32 ± 5	31 ± 4	32 ± 5
Marfan syndrome	14 (70.0)	3 (60.0)	17 (68.0)
Hypertension	4 (20.0)	3 (60.0)	7 (28.0)
Body mass index, kg/m^2	24 ± 4	23 ± 3	24 ± 4
Aortic root diameter, mm	57 ± 14	44 ± 14	54 ± 15
Aortic regurgitation	19 (95.0)	3 (60.0)	22 (88.0)
Mild	1 (5.0)	2 (40.0)	3 (12.0)
Moderate	8 (40.0)	0	8 (32.0)
Severe	10 (50.0)	1 (20)	11 (44.0)
Gestational age, weeks	29 ± 10	27 ± 10	28 ± 10
Median, weeks	31	22	31
1st trimester	2 (10.0)	0	2 (8.0)
2nd trimester	6 (30.0)	3 (60)	9 (36.0)
3rd trimester	7 (35.0)	2 (40)	9 (36.0)
Postpartum	5 (25.0)	0 (0)	5 (20.0)

[a] Continuous data are shown as mean ± SD or as indicated, and categoric data as number (%).

Surgical repair was performed under cardiopulmonary bypass (CPB) and moderate hypothermia. Surgical procedures included composite graft root replacement (Bentall) in 17 patients (68.0%), ascending aortic replacement in 3 (12.0%), total arch replacement (TAR) in 17 (68.0%), frozen elephant trunk (FET) in 18 (72.0%), and thoracoabdominal aortic replacement and extraanatomic bypass in 1 patient (4.0%) each. The technical details of FET + TAR were described in detail previously [18].

Circulatory arrest was used in 18 patients with a mean nasopharyngeal temperature of 23.1 ± 1.6°C (range, 19.0 to 25.0°C). CPB, aortic cross-clamp, and hypothermic circulatory arrest times in the 17 patients undergoing TAR + FET were 196 ± 41 (range, 132 to 316), 107 ± 27 (range, 65 to 173), and 27 ± 10 (range, 15 to 50) minutes, respectively. In 3 patients with an isolated Bentall procedure, the nasopharyngeal temperature was 31.3 ± 4.0° (range, 27.0 to 35.0°C), and the CPB and aortic cross-clamp times were 69 ± 16 (range, 50 to 81) and 46 ± 8 (range, 37 to 52) minutes, respectively. Thoracic endovascular aortic repair (TEVAR) was performed in 1 patient (4.0%) with TBAD.

Patient Follow-Up

All survivors (mothers and fetuses or neonates) were monitored by clinic visits, letters, or phone calls, and by the referring physician to document survival, reoperation, and adverse events. Patients were recommended to have a CT scan annually to evaluate the aorta and detect complications.

Statistical Analysis

Statistical analysis was performed using SPSS for Windows 16.0 (SPSS Inc, Chicago, IL). Data are expressed as mean ± SD (range) or as number and percentages, as appropriate. Long-term survival was estimated using the Kaplan-Meier method.

Results

Type A Aortic Dissection

DELIVERY FIRST. In 7 of 20 patients (35.0%), cesarean delivery was performed first at 38 ± 2 GWs (range, 34 to 40 GWs). Surgical repair was performed after a mean of 41 days (median, 7; range, 2 to 147 days), including Bentall + TAR + FET in 6 patients (2 of whom also had a mitral valve operation), and ascending aortic replacement + TAR + FET in 1 patient (Table 3).

No fetal deaths occurred (0%). One maternal death (14.3%) occurred in a patient who sustained renal failure after Bentall + TAR + FET and required dialysis. She died of multiorgan failure at 20 days.

SINGLE-STAGE DELIVERY AND AORTIC REPAIR. In 6 of 20 patients (30.0%), cesarean section and aortic repair in one stage was performed at a mean of 31 ± 4 GWs (range, 23 to 35 GWs). Surgical procedures included Bentall + TAR + FET (Fig 2) in 4 patients (1 of whom had a coronary artery graft bypass for dissection of the right coronary artery),

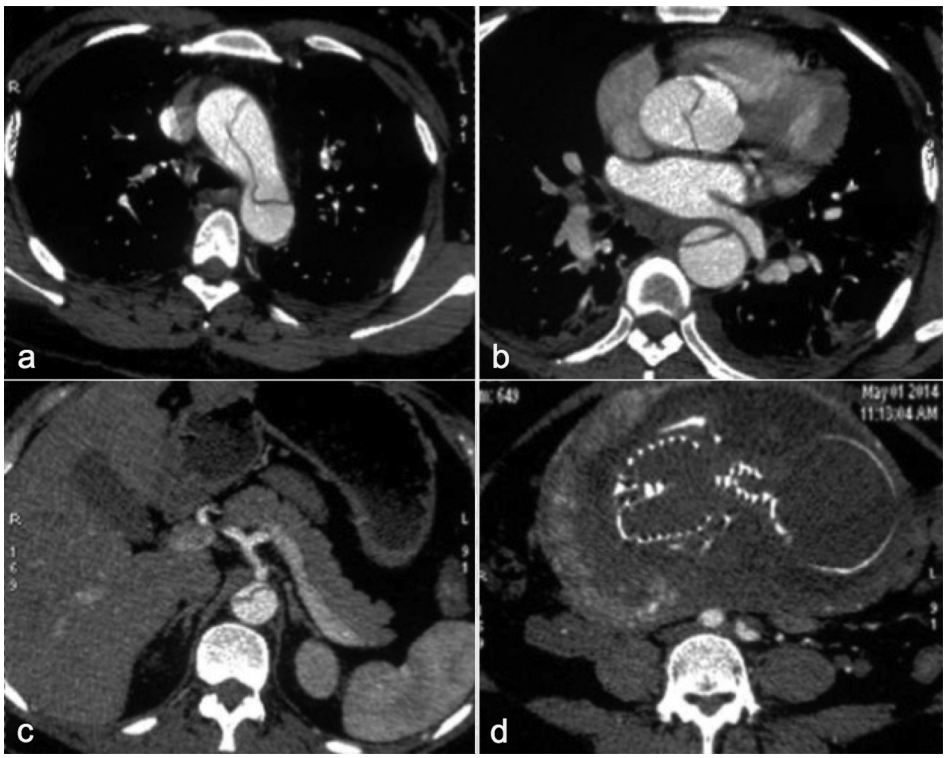

Fig 1. Axial computed tomography images of a 36-year-old woman with acute type A dissection at 26 gestational weeks at the level of the (a) aortic arch, (b) left atrium, (c) celiac trunk, and (d) iliac bifurcation.

isolated Bentall in 1, and ascending aortic and total arch replacement + FET in 1.

Fetal mortality was 33.3% (2 of 6). Two fetuses (of 33 and 23 GWs each) died after cesarean delivery. Maternal mortality was 16.7% (1 of 6). An acute coronary artery graft thrombosis occurred after Bentall + TAR + FET + coronary artery graft bypass in a patient with Marfan syndrome, which necessitated a redo coronary artery graft bypass. Acute renal failure ensued, and she died of multiorgan failure on postoperative day 2.

AORTIC REPAIR FIRST. In 6 of 20 patients (30.0%), because of the earlier gestational age, the dissected aorta was repaired first at 17 ± 7 GWs on average (range, 8 to 27 GWs). Surgical procedures were Bentall + TAR + FET in 3 patients, isolated Bentall in 2, and ascending aortic replacement + TAR + FET in 1.

Maternal mortality was 16.7% (1 of 6), which occurred in a patient after ascending aortic replacement + TAR + FET at 25°C. Her baby (27 GWs) was alive on postoperative day 1. Unfortunately, she died of acute respiratory failure caused by asphyxia at 10 days, leading to fetal death.

After a mean 14 days of hospitalization (median, 11; range, 7 to 28 days), 4 fetuses died intrauterinely, 3 of which were aborted by induced labor. The other 2 fetuses survived after a Bentall procedure done at 19 and 21 GWs and were delivered by cesarean section at 32 and 40 GWs, respectively. Fetal mortality in this group was 66.7% (4 of 6).

MEDICAL MANAGEMENT. An acute TAAD developed at 25 GWs in 1 patient with Marfan syndrome with a chronic TBAD. An immediate urgent operation was recommended, but the patient and her family decided to have medical management first in hope of having single-stage surgical repair and delivery at a later time. Tragically, aortic rupture occurred at 8 days from symptom onset despite aggressive antiimpulse therapy, leading to maternal and fetal death.

Type B Aortic Dissection

DELIVERY FIRST. Among 3 of 5 Marfan patients (60.0%) with TBAD, cesarean delivery was performed in 1 patient at 24 GWs and induced labor in the other 2 at 24 and 23 GWs, respectively. All 3 fetuses died. Surgical repair was performed at 116, 28, and 29 days after delivery, respectively, including thoracoabdominal aortic replacement for a dissecting Crawford extent II aneurysm, FET + left carotid-to-left subclavian bypass for left subclavian involvement, and Bentall + TAR + FET for a large root aneurysm in 1 patient each.

No maternal deaths occurred. Pleural effusion and chest wound infection occurred in the patient with thoracoabdominal aortic repair, which resolved with medical treatment.

AORTIC REPAIR FIRST. In 1 patient (20.0% [1 of 5]), TEVAR was performed first at 38 GWs, and the baby was delivered the next day by cesarean section. The mother and the neonate survived.

MEDICAL MANAGEMENT. One patient sustained an acute TBAD at 37 GWs. After cesarean section, she was transferred to our hospital for further treatment. She was managed with intravenous nitroprusside and an oral

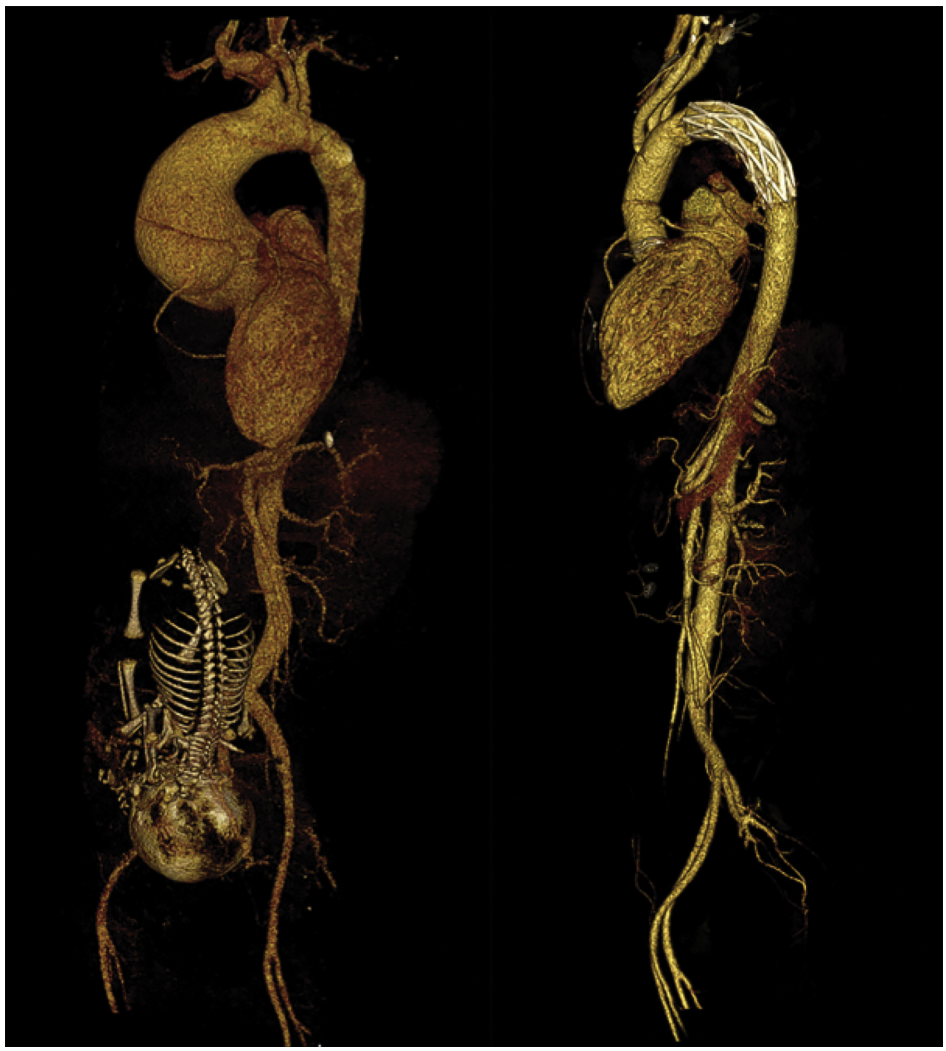

Fig 2. (Left) Sagittal reconstructed computed tomography images of a 27-year-old woman who sustained an acute type A aortic dissection at 29 weeks of gestation. (Right) Maternal and fetal survival was achieved with cesarean delivery, followed by composite root and total arch replacement with frozen elephant trunk implantation in a single stage.

β-blocker (metoprolol). The mother and neonate were discharged uneventfully.

In the 20 mothers with prepartum dissection, the fetal mortality before 28 GWs was 81.8% (9 of 11), which was significantly higher than the 11% (1 of 9) mortality after 28 GWs ($p = 0.005$).

Late Outcomes

Follow-up was complete in 95.3% (20 of 21) averaging 3.2 ± 2.2 years (median, 2.6; range, 0.4 to 8.1 years). One patient was lost to follow-up. Overall maternal survival was 80.0% (95% confidence interval, 58.4% to 91.1%) at 1 year and 3 years and 68.6% (95% confidence interval, 37.5% to 86.5%) at 5 years (Fig 3).

In the TAAD group, late deaths occurred in 3 patients with Marfan syndrome and in 2 fetuses/neonates. The first patient died of over anticoagulation at 3 months after Bentall + TAR + FET + mitral valve replacement. In the second patient, a CT scan detected no aortic complications after a composite graft root replacement, but she died of lymphoma at 4 years. The third patient died of distal aortic rupture at 8 years after Bentall + TAR + FET. No late deaths occurred in the TBAD group.

Secondary intervention was required in 2 patients. The first, a Marfan patient with TAAD, required TEVAR for new entry tear distal to the FET at 1 month postoperatively. A retrograde TAAD developed in the second (non-Marfan) patient with a TBAD at 16 months after TEVAR, which was successfully managed with TAR + FET.

Comment

Although rare in occurrence, an association between pregnancy and aortic dissection was described as early as in the 1880s [19]. According to a review by Yuan [17], approximately 180 cases have been reported in the English literature since Schnitker and Bayer [6] published the first review on aortic dissection during pregnancy in 1944. The rarity and limited experience make it difficult to determine detailed guidelines for the diagnosis and management of this complex clinical scenario [20, 21],

Table 2. Management Strategies

Variables	Aortic Dissection		
	Type A No. (%)	Type B No. (%)	Total No. (%)
Patients	20 (80)	5 (20)	25 (100)
Management strategies			
Surgical	19 (95.0)	3 (60)	22 (88.0)
Endovascular	0	1 (20)	1 (4.0)
Medical	1 (5.0)	1 (20)	2 (8.0)
Surgical procedures			
Composite graft root replacement	16 (80.0)	1 (20)	17 (68.0)
Ascending aortic replacement	3 (15.0)	0	3 (12.0)
Total arch replacement	16 (80.0)	1 (20)	17 (68.0)
Frozen elephant trunk	16 (80.0)	2 (40)	18 (72.0)
Thoracoabdominal aortic replacement	0	1 (20)	1 (4.0)
Left carotid–left subclavian bypass	0	1 (20)	1 (4.0)
Timing of aortic repair and delivery			
Delivery before aortic repair			
In 2 stages	7 (35.0)	3 (60)	10 (40.0)
In 1 stage	6 (30.0)	0 (0)	6 (24.0)
Aortic repair before delivery	6 (30.0)	1 (20)	7 (28.0)

which prompted us to report our experience with an emphasis on the etiology, management strategies, and clinical outcomes in 25 such patients during a 17-year period.

Aortic dissection typically occurs in the third trimester of pregnancy or during the early postpartum period [22]. Although connective tissue disorders, aortic root diameter of 40 mm or more, hypertension, bicuspid aortic valve, coarctation, and so on can predispose pregnant women to the occurrence of aortic dissection [17], some believe that pregnancy alone, with no underlying causes, is an independent risk factor for aortic dissection [23].

The demographic features of patients in this series are similar. In 56% of the patients, aortic dissection occurred in the third trimester and in the early postpartum period. Marfan syndrome was the predominant risk factor, as seen in 68% of the patients. Another finding is that the diameter of the aortic root measured 54 ± 15 mm for the entire group, which shows the risks of aortic dissection associated with an aortic root diameter of 40 mm or more in pregnant women [14, 17]. The incidence of hypertension was 28% in this cohort, in contrast to 76.6% (3,247 of 4,428) found in the International Registry of Acute Aortic Dissection database [24]. The relatively low prevalence of hypertension in this cohort might imply that distinct effects of pregnancy are likely and that pregnancy itself is an independent risk factor for aortic dissection [4, 23].

Our experience with surgical treatment of aortic dissection in pregnancy dated back to 1998 when the senior author (L.Z.S.) successfully performed an emergency Bentall procedure in a 38-year-old woman with Marfan syndrome who sustained an acute type II aortic dissection at 32 GWs. A cesarean section was performed in the same stage and delivered a stillborn fetus, which was most likely caused by intrauterine asphyxia before the induction of anesthesia.

During the past 17 years, we have managed 25 pregnant women with aortic dissection, and our treatment algorithm evolved considerably toward the goal of saving 2 lives. Because the experimental studies from Hanley's group [25] show that CPB may result in lower placental flow and pressure, which are worsened by hypothermia and lead to impaired placental perfusion, our current perfusion strategies include using warmer temperatures (normothermia or mild hypothermia) during CPB, minimizing CPB times, avoiding circulatory arrest before 28 GWs, and maintaining a high flow rate (>2.4 L/m^2 per minute) and mean arterial pressures exceeding 70 mm Hg [26]. Over time, cold crystal cardioplegia was replaced by cold blood cardioplegia for better myocardial protection.

In 2009 our team moved to the current center where we can collaborate closely and efficiently with a group of obstetricians with expertise in cardiovascular problems in pregnant women who offer high-quality care for both the mother and her fetus/neonate and allow for making structured management decisions. Now the uterus is routinely managed by insertion of a Cook balloon after cesarean delivery instead of a hysterectomy. Survival of fetuses before 28 GWs has been achieved after cesarean delivery.

Although a CT angiogram is usually the preferred tool for the diagnosis of aortic dissection, considering the radiation damage to the unborn child, more recently we have been trying to use a reliable echocardiogram as an early diagnostic tool for such patients. Because of these improvements, we have achieved an overall early survival of 84% for the mothers and 60% for the fetuses/neonates, and an overall long-term maternal survival of 80% at 3 years and 68.6% at 5 years. On the basis of these results, we speculate that our algorithm, although slightly different from others [13, 14, 17], might be helpful in the management of patients with aortic dissection during pregnancy.

The management decision should be based on the gestational age and the type of dissection. For TAAD occurring before 28 GWs, urgent surgical repair with aggressive fetal monitoring or abortion is preferred. Because of the high risks of fetal death during hypothermic circulatory arrest or CPB [27], maternal survival should be prioritized over fetal concerns. When dissection occurs after 28 GWs, urgent cesarean section followed by aortic repair appears to offer the best chance for maternal and fetal survival. For TBAD, medical therapy or TEVAR is preferred unless open surgical repair is mandated by malperfusion or aortic rupture.

With respect to surgical procedures, composite graft root replacement (Bentall) without circulatory arrest is preferred for dissections located in the root or ascending aorta, and FET and total arch replacement is preferred for dissections involving the arch or descending aorta,

Table 3. Early and Late Outcomes

	Entire Cohort			Type A Aortic Dissection			Type B Aortic Dissection		
Variables[a]	Total	Alive	Dead	Subtotal	Alive	Dead	Subtotal	Alive	Dead
Early outcomes									
Maternal survival and mortality	25	21 (84.0)	4 (16.0)	20	16 (80.0)	4 (20.0)	5	5 (100)	0
Delivery before aortic repair in 2 stages	10	9 (90.0)	1 (10.0)	7	6 (85.7)	1 (14.3)	3	3 (100)	0
Single-stage delivery and aortic repair	6	5 (83.3)	1 (16.7)	6	5 (83.3)	1 (16.7)	0	NA	NA
Surgical repair before delivery	6	5 (83.3)	1 (16.7)	6	5 (83.3)	1 (16.7)	0	NA	NA
TEVAR before delivery	1	1 (100)	0	0	NA	NA	1	1 (100)	0
Medical therapy	2	1 (100)	0	1	0	1 (100)	1	1 (100)	0
Fetal/neonatal survival and mortality	25	15 (60.0)	10 (40.0)	20	13 (65.0)	7 (35.0)	5	2 (40.0)	3 (60.0)
Delivery before aortic repair in 2 stages	10	7 (70.0)	3 (30.0)	7	7 (100)	0	3	0	3 (100)
Single-stage delivery and aortic repair	6	4 (66.7)	2 (33.3)	6	4 (66.7)	2 (33.3)	0	NA	NA
Surgical repair before delivery	6	2 (33.3)	4 (66.7)	6	2 (33.3)	4 (66.7)	0	NA	NA
TEVAR before delivery	1	1 (100)	0	0	NA	NA	1	1 (100)	0
Medical therapy	2	1 (50.0)	1 (50.0)	1	0	1 (100)	1	1 (100)	0
Fetal/neonatal weight, kg	15	2.2 ± 2.0	NA	13	2.4 ± 1.0	NA	2	2.2 ± 2.0	NA
	Total	Yes	No	Subtotal	Yes	No	Subtotal	Yes	No
Late outcomes									
Lost to follow-up	21	1 (4.8)	20 (95.2)	15	1 (6.7)	14 (93.3)	5	0	5 (100)
Maternal survival	20	17 (85.0)	3 (15.0)	15	12 (80.0)	3 (20.0)	5	5 (100)	0
Fetal/neonatal survival	15	13 (86.7)	2 (13.3)	13	11 (84.6)	2 (15.4)	2	2 (100)	0
Aortic reintervention	20	18 (90.0)	2 (10.0)	15	14 (93.3)	1 (6.7)	5	1 (20.0)	4 (80.0)

[a] Continuous data are shown as mean ± SD and categoric data as number (%).

NA = not applicable; TEVAR = thoracic endovascular aortic repair.

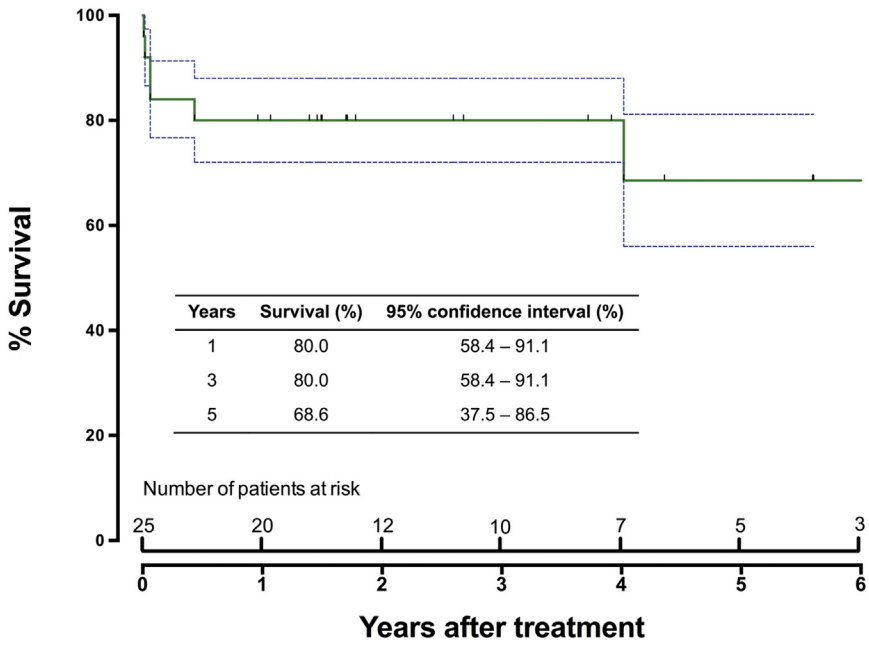

Fig 3. Overall maternal survival (green line) after management of aortic dissection in pregnancy. The blue lines show the 95% confidence interval.

especially in younger patients or those with Marfan syndrome. Other centers may follow specific procedures that are most comfortable for the surgical team in their general aortic environment.

This study also highlights the poor prognosis of women with Marfan syndrome when aortic dissection occurs in pregnancy. Two of the 4 early deaths (50%) occurred in Marfan patients, as did 3 of 3 late deaths (100%) and 1 of 2 late reinterventions (50%). Aortic dissection may develop in women with Marfan syndrome in all 3 trimesters. Even after aggressive surgical repair, they are still at risk of late death and complications. It is therefore highly recommended that women with Marfan syndrome be counseled about the risk of aortic dissection and the heritable nature of the disease before pregnancy [28].

This study has several limitations. It is retrospective in nature and has a small sample size. Follow-up was incomplete and of short duration.

Because of the peculiarities of China's health care system, few centers in China nowadays have established aortic surgical programs, and even fewer have comprehensive on-call arrangements for multidisciplinary aortic teams capable of managing acute aortic dissection as an emergency. To alleviate this desperate situation, we set up a collaborative mechanism whereby certain aortic surgeons, anesthesiologists, and perfusionists with expertise may travel urgently to local or peripheral hospitals to operate on acute unstable patients who cannot be transferred. This led to, in part, the delay in operative repair for some patients with TAAD and explained why 10 patients from 9 other hospitals (Supplementary Table 2) were included this study, all of whom were operated on by the first author (J.M.Z.). Although the same protocols for the operation, anesthesia, perfusion, and intensive care were used to manage patients in this cohort, the effect of institutional differences on treatment outcomes cannot be overlooked and might be an indirect factor that affected the clinical outcomes, especially given the slight difference that may exist with the cardiac and obstetric nursing teams and postnatal care at different hospitals. As a result, detailed information on fetal outcomes, such as the Apgar score of the neonates, was incomplete. All of these limitations make it difficult to make any conclusive recommendations.

In conclusion, Marfan syndrome was the leading etiology of aortic dissection in pregnancy. Management should be based on the type of dissection and gestational age. In this series, for TAAD occurring after 28 GWs, cesarean section followed by surgical repair could achieve maternal and fetal survival adequately; before 28 GWs, maternal survival should be prioritized over fetus concerns. For patients with TBAD, nonsurgical management is preferred, if possible.

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