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REVIEW ARTICLE

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Pregnancy-related acute aortic dissection in Marfan syndrome: A review of the literature

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Abstract

A well-established association exists between acute aortic dissection and pregnancy, particularly in women with Marfan syndrome. However, there is debate regarding appropriate management guidelines. In particular, there are differing opinions regarding when prophylactic aortic root repair should be recommended as well as the efficacy of beta blockers in this clinical scenario. The current study evaluated 10 years of published literature (2005-2015) in the PubMed/Medline database. Fifty articles, describing 72 cases of women who presented with aortic dissection in the antepartum or postpartum period were identified. Comparisons on demographic variables and clinical outcomes between cases of women with Marfan syndrome ($n = 36$) and without Marfan syndrome ($n = 36$) were conducted. There were no significant differences in demographics (age, gravidity, parity) between the Marfan and non-Marfan cases. Marfan patients presented with antepartum dissections significantly earlier in pregnancy than those without Marfan syndrome ($P = .002$). However, there were no significant difference between the 2 groups in maternal mortality, fetal mortality, or obstetric outcomes (mode of delivery and gestational age at delivery). Eight cases described events in Marfan women with an aortic root diameter $\leq$40 mm. Six events occurred in Marfan women who were managed with beta blockers. Current guidelines rely on aortic root diameter for stratification of Marfan women into risk categories, but we identified several cases that would be missed by these guidelines. Specifically, the existing literature suggest that women with Marfan syndrome should take precautions throughout pregnancy, rather than the third trimester.

KEYWORDS

Aortic dissection, aortic root, beta blocker, congenital heart disease, Marfan syndrome, pregnancy

1 | INTRODUCTION

Pregnancy is an inherently hypervolemic and hyperdynamic cardiovascular state.[1] Physiologic changes during pregnancy include increases in maternal blood volume, heart rate, stroke volume, and cardiac output. Pregnancy-related elevations in estrogen and progesterone may decrease the amount of mucopolysaccharides and elastic fibers in the tunica media of the aorta, resulting in an average increase in aortic root diameter of 1 mm in healthy patients.[2–4] These pregnancy-related increases in aortic root diameter appear to be more pronounced in patients with Marfan syndrome (MFS), with an average increase of 3 mm in diameter.[5] Because of the physiologic changes associated with pregnancy, cardiovascular disease complicates 1%-3% of pregnancies and is the second most common cause of maternal mortality.[6] Nearly half of these deaths are due to aortic dissection, and half of all aortic dissections in women younger than 40 years are related to pregancy.[7] Schnitker first described the relationship between aortic dissection and pregnancy.[8] This association persists throughout the literature, but it has been suggested that it may be an artefact of selective reporting.[9,10]

The composite risk from Marfan syndrome and pregnancy increases the likelihood of acute aortic dissection (AAD) in these patients.[11] Current recommendations from the World Health Organization (WHO) classify women with MFS into 4 risk categories based on the extent of their cardiovascular condition (Table 1).[12] The WHO classifications are primarily based on aortic root measurements, as this is believed to be the greatest risk factor for the development of AAD. However, even with a preconception aortic diameter less than 40 mm,

TABLE 1 WHO pregnancy classifications for women with Marfan syndrome[12]

Risk category	Qualifications	Treatment recommendations[a]
WHO I	Mitral valve prolapse	Echocardiography every 4-8 weeks
WHO II	Aortic root diameter <40 mm	Vaginal delivery unless otherwise indicated
		Echocardiography every 4-8 weeks
WHO III	Aortic root diameter 40-45 mm	Consider cesarean delivery
		Prophylactic surgery prior to conception
WHO IV	Aortic root diameter >45 mm	Consider prophylactic surgery during pregnancy

[a]All women with MFS should be advised on the risks of AAD and transmission to children.

there is a 1% risk of dissection during pregnancy.[13] This risk is increased to 10% with an aortic diameter >40 mm.[14] The American College of Cardiology recommends that women who are contemplating pregnancy should undergo prophylactic aortic repair if the diameter of their aortic root exceeds 40 mm (Level C).[15] The Canadian and European guidelines recommend that women who are contemplating pregnancy undergo prophylactic aortic repair if their aortic root diameter exceeds 45 mm.[12,16]

Delivery guidelines for women with MFS are based on aortic root diameter. Women with an aortic root <40 mm are advised toward vaginal delivery.[17,18] Women with an aortic root diameter >45 mm should be delivered by cesarean section. The evidence is less clear for delivery modality in women with an aortic diameter of 40-45 mm. The WHO guidelines recommend that clinicians consider cesarean for these women, but other studies suggest that vaginal delivery with an expedited second stage of labor can be used.[7,15,19] Women with MFS are at an increased risk of dissection for at least 6 months during the postpartum period, but some studies suggest that this risk may remain elevated for at least 1 year after delivery.[18]

Several previous studies have evaluated the incidence of AAD during pregnancy in women with MFS (Table 2).[5,13,18,20–28] In total, these 12 studies evaluated 1271 pregnancies in at least 832 women with MFS. Thirty-nine of these pregnancies were complicated by an AAD. The composite incidence of aortic dissection in these studies constitutes a risk of 3.07% for AAD during a pregnancy for a woman with Marfan syndrome. This value does not consider factors such as beta blocker usage, prophylactic aortic repair, and aortic root size. Additionally, many women with MFS and a dilated aortic root are advised to avoid pregnancy.[12,13,18] For these reasons, the true risk of AAD during pregnancy for women with MFS may be underestimated, especially for those with a dilated aortic root.

We will analyze the published literature that describes cases of AAD in the antepartum and postpartum period. Published cases of pregnancy-related AAD will be compared between cases with and without a diagnosis of Marfan syndrome. Our aim is to evaluate the demographics, presentation, and outcomes of these cases to elucidate the factors that may contribute to the development of aortic dissection during pregnancy in women with MFS. From this information, we hope to identify the key factors that obstetricians and cardiologists should consider when treating pregnant Marfan women.

TABLE 2 The incidence of AAD in published population-based studies on pregnancy in Marfan syndrome

	Year	Marfan cases (*n*)	Pregnancies (*n*)	Aortic dissections (*n*)
Pyeritz et al.[13]	1981	26	105	0
Rossiter et al.[20]	1995	21	45	2
Lipscomb et al.[21]	1997	36	91	4
Lind and Wallenburg[22]	2001	44	78	5
Meijboom et al.[18]	2005	23	33	1
Meijboom et al.[23]	2006	122	142	1
Pacini et al.[24]	2009	85	160	7
Katsuragi et al.[25]	2011	28	28	11
Donnelly et al.[5]	2012	69	199	0
Omnes et al.[26]	2013	18	22	1
Curry et al.[27]	2014	21	29	1
Hassan et al.[28]	2015	not given	339	6
	Total	**832**	**1271**	**39**

2 | METHODS

We searched the PubMed/Medline database (January 2005-November 2015) for English-language articles containing combinations of the following terms: "aortic dissection"; "Marfan syndrome"; "aortic aneurysm"; and "pregnancy." The search for "Marfan syndrome" and "pregnancy" yielded a total of 143 results (63 case reports); the search for "aortic dissection" and "pregnancy" yielded 214 results (76 case reports); and the search for "aortic aneurysm" and "pregnancy" yielded 222 results (109 case reports). A total of 248 case reports and case series were retrieved. Of these studies, 198 articles were ruled out based on title and/or abstract or were repeated across the database searches. Case reports and case series were included in our analysis if the patient had an acute aortic dissection during pregnancy or during the first 12 months postpartum. The 12-month postpartum cutoff was chosen because it is in accordance with other similar studies.[25] Only cases that resulted in a dissection were included in this analysis.

We identified 50 articles that fit our inclusion criteria (41 solitary cases and 9 case series).[29–79] The 50 articles yielded a total of 72 patients who met the inclusion criteria (AAD during pregnancy or within 1 year postpartum). When available, data for the following parameters were extracted from the articles: maternal age, gravidity, parity, Marfan syndrome diagnosis, aortic root measurements, Stanford type, weeks gestational age (WGA) at dissection, mode of delivery, WGA at delivery, maternal outcome, fetal outcome, surgical management, and medical management.

2.1 | Statistical analysis

Continuous variables (age, gravidity, parity, WGA at dissection, WGA at delivery, aortic root measurements) are reported using descriptive statistics (mean ± standard deviation or median and range, as appropriate) and between group comparisons of the MFS and non-MFS patients were conducted with an independent *T*-test. Associations of dichotomous variables (Stanford classification, delivery method, maternal outcome, fetal outcome) between the MFS and non-MFS patients were conducted with a Chi-square test (X^2) and reported as frequency and percentage. For this study, all statistical tests were 2-tailed and statistical significance was defined as a *P*-value $< .05$. Statistical tests were conducted in SPSS 22.0.[80]

3 | RESULTS

Of the 72 patients included in this study, 36 patients were diagnosed with MFS and 36 of the patients were not diagnosed with MFS. The cases were stratified into "MFS" and "non-MFS" groups based on this variable. The mean age ± standard deviation (SD) of the sample was 32.21 ± 4.70 (32.50 ± 4.46 in the MFS group, 31.92 ± 4.92 in the non-MFS group, $P = .605$). The average parity of the entire sample was 0.98 ± 1.03 (1.04 ± 1.21 in the MFS group, 0.90 ± 0.77 in the non-MFS group; $P = .6597$). The average gravidity of the entire sample was 2.18 ± 1.23 (2.42 ± 1.38 in the MFS group, 1.90 ± 0.94 in the non-MFS group; $P = .1729$). Seventeen of the patients were nulliparous (10 in the MFS group, 7 in the non-MFS group). Obstetric history could not be obtained for 28 of the patients. General demographic data can be found in Table 3.

TABLE 3 General demographic data from the case reports analyzed in this review

	MFS cases (*n* = 36)	Non-MFS cases (*n* = 36)	All cases (*n* = 72)	*P* value
Patient age (years)				
Mean ± SD	32.50 ± 4.46	31.92 ± 4.92	32.21 ± 4.70	.6047
Median	32.5	32	32	
Range	23 - 41	22 - 40	22 - 41	
Obstetric history[a]				
Gravidity				
Mean ± SD	2.42 ± 1.38	1.90 ± 0.94	2.18 ± 1.23	.1729
Median	2	2	2	
Parity				
Mean ± SD	1.04 ± 1.21	0.90 ± 0.77	0.98 ± 1.03	.6597
Median	1	1	1	
Nullipara (*n*)	10	7	17	

[a]Obstetric history was not reported for 12 of the MFS cases and 16 of the non-MFS cases.

Most of the acute aortic events occurred in the third trimester ($n = 40$, 55.6%) and the postpartum period ($n = 21$, 29.2%) (Table 4). Several patients presented during the second trimester ($n = 9$, 12.5%). Few presented with a dissection in the first trimester ($n = 2$, 2.8%). The timing of presentation differed between the MFS and non-MFS groups. In the MFS group, half of the patients presented in the third trimester ($n = 18$, 50.0%), and an equal number of patients presented with second trimester and postpartum dissections ($n = 8$, 22.2%). Two patients in the MFS group presented with a dissection in the first trimester ($n = 2$, 5.6%). The earliest and latest events in the MFS group were 7 WGA and 7 months postpartum, respectively. In the non-MFS group, most patients presented during the third trimester (22, 61.1%) and postpartum ($n = 13$, 36.1%). One patient in this group presented with a dissection during the late second trimester, at 26 WGA ($n = 1$, 2.8%). The latest event in the non-MFS group was at 2 months postpartum.

For all antepartum events, the mean ± SD WGA at dissection for the MFS ($n = 28$) and non-MFS ($n = 23$) groups were 27.71 ± 8.00 and 33.74 ± 3.72, respectively ($P = .002$). This represents a statistically significant difference in the timing of antepartum aortic dissections between pregnant women with MFS and without MFS. The median WGA at dissection for antepartum events in the MFS group was 29.5 (range 7-38); the median WGA for the non-MFS group was 33 (range 26-41 WGA).

Postpartum events were analyzed in a similar manner. The mean ± SD days after delivery for postpartum dissections in the MFS group ($n = 8$) was 44 ± 78.50; in the non-MFS group ($n = 13$), these values were 11.23 ± 14.78, $P = .280$). The large standard deviation in the MFS group for postpartum dissections can be attributed to 2 outliers of dissections at 7-months postpartum and 4-months postpartum.[30,54] The median values for days after delivery for presentation with a postpartum dissection were 5.5 days (range 1 day-7 months) in the MFS group and 7 days (range 1 day-2 months) in the non-MFS group.

TABLE 4 Aortic parameters from case reports

	MFS cases ($n = 36$)	Non-MFS cases ($n = 36$)	All cases ($n = 72$)	*P* value	Odds ratio (confidence interval)
Stanford classifications	*n* (%)	*n* (%)	*n* (%)		
Type A	26 (72.22)	27 (75.00)	53 (73.61)	.789	0.867(0.303-2.475)
Type B	10 (27.78)	9 (25.00)	19 (26.39)		
Aortic root diameter measurements (mm)					
Number of values (n)[a]	22	6	28	.140	
Mean ± SD (mm)	51.72 ± 15.66	45.17 ± 5.79	50.32 ± 14.39		
Median (mm) and range	51.5 (28–85)	43 (40-55)	48 (28-85)		
Dissection timing					
Antepartum events (n)	28	23	51		
mean ± SD	27.71 ± 8.00	33.74 ± 3.72	30.43 ± 7.08	.002	
Median (WGA) and range	29.5 (7-38)	33 (26-41)	32 (7-41)		
Postpartum events (n)	8	13	21		
Mean ± SD (days)	44 ± 78.50[b]	11.23 ± 14.78	23.71 ± 50.64	.280	
Median (days)	5.5	7	7		
Range	1 d-7 mo PP	1 d-2 mo PP	1 d-7 mo PP		
Dissection timing	*n* (%)	*n* (%)	*n* (%)		
1st trimester[c] (≤12 WGA)	2 (5.56)	0 (0.00)	2 (2.78)		
Type A	2	0	2 (100)		
Type B	0	0	0 (0)		
2nd trimester (13-27 WGA)	8 (22.22)	1 (2.77)	9 (12.5)		
Type A	5	0	5 (55.56)	.236	1.333 (0.757-2.348)
Type B	3	1	4 (44.44)		
3rd trimester (28 WGA-term)	18 (50.00)	22 (61.11)	40 (55.56)		
Type A	13	19	32 (80)	.266	0.411 (0.083-2.025)
Type B	5	3	8 (20)		
Postpartum (delivery-12 mo)	8 (22.22)	13 (36.11)	21 (29.17)		
Type A	6	8	14 (66.67)	.525	1.875 (0.266-13.202)
Type B	2	5	7 (33.33)		

[a]AoR diameter measurements were not reported in 14 of the MFS cases and 30 of the non-MFS cases.
[b]Large SD attributable to cases of AoD at 7 months postpartum[54] and 4 months postpartum.[30]
[c]Chi-square analysis was determined to be inappropriate for this data.
WGA, weeks gestational age; MFS, Marfan syndrome.

Stanford classifications were used to further characterize the aortic events. Most dissections were Stanford Type A ($n = 53$, 73.6%), with a smaller portion of Stanford Type B dissections ($n = 19$, 26.4%). The breakdown of dissection types was similar between the MFS and non-MFS groups. The distribution of Type A and Type B dissections did not differ significantly between the MFS and non-MFS groups with each trimester evaluated separately. Table 4 depicts the *P* values and odds ratios for these analyses.

Aortic root diameters were reported for 22 (61.11%) of the MFS cases and 6 (16.67%) of the non-MFS cases. Most of these measurements were taken at the time of the acute aortic event. Only one of the cases reported serial aortic root measurements over the course of the pregnancy.[38] The mean aortic root diameters for MFS cases and non-MFS cases were 51.7 ± 15.7 mm and 45.2 ± 5.8 mm, respectively ($P = .140$). The aortic root diameters in the MFS group range from 28 to 85 mm with a median of 51.5 mm. This is a much wider range than was seen in the non-MFS group. The non-MFS cases range from 40 to

TABLE 5 Obstetric outcomes and mortality data from case reports

	MFS cases ($n = 36$)	Non-MFS cases ($n = 36$)	All cases ($n = 72$)	*P* value	Odds ratio
Maternal outcomes[a]	*n* (%)	*n* (%)	*n* (%)		
Alive	27 (79.41)	28 (77.78)	55 (78.57)	.868	1.102 (0.351-3.459)
Dead	7 (20.59)	8 (22.22)	15 (21.43)		
Fetal outcomes[b]	*n* (%)	*n* (%)	*n* (%)		
Alive	30 (88.24)	30 (88.24)	60 (88.24)	1.000	1.000 (0.229-4.373)
Dead	4 (11.76)	4 (11.76)	8 (11.76)		
Obstetric outcomes					
Delivery method[c]	*n* (%)	*n* (%)	*n* (%)		
Cesarean	26 (72.22)	28 (80.00)	54 (76.05)	.191	
Vaginal	8 (22.22)	5 (14.29)	13 (18.31)		
Abortion	1 (2.78)	0 (0.00)	1 (1.41)		
IUFD	1 (2.78)	2 (5.71)	3 (4.23)		
WGA at delivery[d]					
Mean ± SD	32.44 ± 4.03	33.5 ± 3.86	32.94 ± 3.95	.364	
Median	33	33	33		
Range	22-39	26-41	22-41		
Fetus in utero	2	2	4		

[a]Maternal outcomes were not reported for 2 of the MFS cases.
[b]Fetal outcomes were not reported for 2 of the MFS cases and 2 of the non-MFS cases.
[c]Delivery method was not reported for 1 of the non-MFS cases.
[d]WGA at delivery was not reported for 7 of the MFS cases and 12 of the non-MFS cases.

55 mm with a median of 43 mm. In the MFS group, 10 aortic root diameter measurements were less than 45 mm and 8 aortic root measurements were ≤40 mm. In the non-MFS group, 3 aortic root diameter measurements were less than 45 mm (Table 4).

Only 6 of the MFS cases reported beta-blocker therapy during the pregnancy.[35,44,47,52,54,79] None of the cases with non-MFS patients reported beta-blocker therapy.

We analyzed several obstetric outcomes (delivery method and WGA at delivery) for the pregnancies described in the literature (Table 5). Delivery methods were similar between the MFS and non-MFS groups, with most infants delivered by cesarean ($n = 54$, 76%). There was no significant difference in delivery mode between the 2 groups ($P = .191$). The mean ± SD WGA at delivery for the MFS group was 32.44 ± 4.03 with a median of 33 weeks (range 22-39). For the non-MFS group, mean ± SD WGA at delivery was 33.5 ± 3.86 with a median of 33 weeks (range 26-41). There was no significant difference in WGA at delivery between the MFS and non-MFS groups ($P = .364$).

3.1 | Maternal and fetal mortality

The cases reported a total of 55 positive maternal outcomes and 15 maternal deaths, representing a total maternal mortality of 21.43% (Table 5). Maternal outcomes could not be ascertained for 2 of the patients. There were 7 maternal deaths in the MFS case reports and 8 maternal deaths in the non-MFS case reports. Twelve of these deaths were associated with Type A dissections and three were associated with Type B dissections. The most common causes of death referenced in these cases were low cardiac output syndrome ($n = 6$), multiorgan failure ($n = 2$), and sudden cardiac death from the AAD ($n = 3$). The entire sample showed 60 positive fetal outcomes and 8 fetal deaths, representing a total fetal mortality of 11.76% (Table 5). There were 30 positive fetal outcomes and 4 fetal deaths in each of the MFS and non-MFS groups. Pagni et al. reported on a patient with a positive fetal outcome in a twin pregnancy.[36] This review primarily focuses on maternal pathology, so this was analyzed as one positive fetal outcome. Fetal outcomes could not be ascertained for 4 of the case reports. There was no significant difference in maternal mortality ($P = .868$) or fetal mortality ($P = 1.000$).

4 | DISCUSSION

To our knowledge, this is the first literature review focusing specifically on published cases of pregnancy-associated AAD in Marfan patients. Our review and analysis of the literature revealed several key findings. Perhaps the most significant finding was that women with Marfan syndrome presented with aortic dissection significantly earlier in pregnancy than those without MFS. An additional item of interest from this review were the reported aortic root measurements. Out of the 22

MFS cases that reported aortic root diameter, 10 of these cases reported aortic root diameters <45 mm at the time of dissection, and 8 reported aortic root diameters ≤40 mm. Based on the current guidelines, these women may have "slipped through the cracks" and failed to receive recommendations for prophylactic aortic root repair. Finally, we will discuss our results regarding the use of beta blockers in these cases. Following is a detailed discussion on each of these findings.

4.1 | Early diagnosis

Diagnosis of MFS should be established as early as possible. This can be challenging because some women with MFS may not display obvious Marfanoid features. Furthermore, an estimated 25% of MFS cases arise from de novo mutations. The lack of family history coupled with the subtle presentation can make these patients particularly challenging to diagnose. We identified 4 case reports of pregnancy-related AAD in patients with undiagnosed MFS. In these cases, the diagnosis of Marfan syndrome was made by genetic testing after the patient presented with a dissection.[41,42,44,45] Chang, Katsuragi, and Lichtman reported cases where no Ghent features were identified other than the acute aortic event. This suggests that additional criteria may be needed for the diagnosis of Marfan syndrome. With earlier diagnosis, it is possible that an acute aortic event may have been avoided if appropriate precautions were taken throughout these pregnancies, such as beta blockade, prophylactic aortic root repair, and serial aortic root measurements. Unfortunately, the literature does not suggest any specific features that may be used as a screening tool for these undiagnosed cases of MFS in obstetric patients.

Early MFS diagnosis is especially important because aortic dissection can occur early in pregnancy in these patients. The earliest event described in the literature was at 7 WGA. It is unclear from these cases, if pregnancy was a significant component in the development of aortic dissection or if the event was primarily driven by an aorta that was susceptible to dissection prior to conception. Furthermore, these cases did not report if these patients had a family history of AAD. These early events are significant because the current literature on pregnancy-related AAD focuses on the predominance of acute aortic events in the third trimester and postpartum. Although most of the cases identified in this study do support this timeline, the evidence for earlier dissections in MFS patients should be considered. We suggest that Marfan patients and their families should be taught to recognize the early symptoms of aortic dissection at their first prenatal appointment or prior to conception.

The timing of aortic dissection in pregnancy is also important to consider from a management perspective. If a patient presents with an aortic dissection later in pregnancy, it is possible to perform an emergency cesarean section followed by an aortic repair. In women who present with AAD before fetal viability, the patient and physician must decide between termination of the pregnancy prior to surgical repair and surgical repair with the fetus in utero, which presents considerable risks to both the mother and the fetus.[81] Because women with MFS present with AAD significantly earlier than those without MFS, more of these women may be required to make this difficult decision. However, it is important to note that our evaluation did not find a significant difference in maternal mortality, fetal mortality, or obstetric outcomes (WGA at delivery or mode of delivery) between the MFS cases and the cases of women without MFS.

In addition to early diagnosis of MFS, the importance of early identification of aortic dilatation is critical, especially when early intervention can be performed to reduce the risk of AAD. Our literature search revealed 5 cases of aneurysms that were successfully repaired before the possibility of dissection. Sato et al. describes a MFS patient who presented with an aortic aneurysm at 15 WGA, which was repaired with fetus in utero at the time of presentation. The procedure was successful, and a healthy infant was delivered at 37 WGA by cesarean. The mother did not present with an aortic dissection at any time during pregnancy or postpartum.[82] Gama et al. describes a case in which an aortic aneurysm (69 mm) was detected at 10 WGA in a patient without Marfan syndrome. The aneurysm was repaired with fetus in utero at the time of diagnosis. A healthy infant was delivered by cesarean section at 38 WGA with positive maternal and fetal outcomes.[83] Tutarel et al. and Volach et al. both describe cases in which an aortic root replacement was performed in MFS patients prior to conception with no cardiovascular complications during pregnancy and positive maternal and fetal outcomes.[84,85] These cases suggest that early identification of a dilated aortic root and subsequent repair can protect the maternal prognosis without sacrificing the prognosis of the fetus.

4.2 | Aortic root measurements

The literature indicates that some women with MFS who present with an aortic dissection during pregnancy may not be appropriately identified by the current guidelines. The current recommendations for women with MFS include prophylactic aortic repair before conception if the aortic root diameter is ≥40 mm (US guidelines) or ≥45 mm (Canadian and European guidelines). In the MFS group, 22 cases reported aortic root measurements. Of these 22 cases, 10 women had an aortic root diameter <45 mm; 8 of these women had an aortic root diameter ≤40 mm. The minimum aortic root diameter was 28 mm at the time of presentation. This data suggests against the current notion that pregnancy is relatively low risk in MFS women with an aortic root diameter ≤40 mm. Furthermore, these women would not have been identified as "high risk" based on the current guidelines and would not have received recommendations to either avoid pregnancy or receive prophylactic aortic root repair before conception.

Serial aortic root measurements throughout pregnancy were only provided by one case identified in this study. We believe that the lack of data regarding the growth rate of the aortic root throughout these pregnancies suggests a deficit in the literature that should be further investigated. Meijboom et al. performed a prospective study that found differing growth rates in the aortic roots of a cohort of Marfan women during pregnancy.[18] We would be interested to see if patients with "fast growth" are more likely to experience an acute aortic event during pregnancy, regardless of preconception aortic root size. Furthermore, we believe that there is a need for evaluation of the factors that

contribute to differential aortic growth rates in some Marfan individuals during pregnancy.

We understand that a publication bias may exist that underemphasizes the importance of aortic root diameter in the pathogenesis of pregnancy-related AAD in women with Marfan syndrome. However, the cases identified in our search indicate that women with Marfan syndrome and an aortic root diameter ≤40 mm may be at a higher risk for AAD than previously believed. At this point, we believe that it would be reasonable to recommend more frequent aortic root measurements during pregnancy in women with Marfan syndrome, regardless of aortic root diameter. One consideration could be that all women with Marfan syndrome should all be treated as high risk for dissection and should be under careful surveillance by a multidisciplinary team including cardiologists, obstetricians, and perinatologists.

4.3 | Beta blockers

Beta-blocker therapy has long been considered a mainstay of treatment for patients with Marfan syndrome, but its efficacy has been the subject of continued debate. Additionally, there exists limited evidence regarding the importance of beta blockade during pregnancy.[16] Current recommendations include daily beta-blocker therapy for pregnant women with MFS. In the literature identified in this study, only 6 of the MFS cases reported beta-blocker therapy during the pregnancy.[35,44,47,52,54,79] It is unclear if the other MFS patients were not receiving beta-blocker therapy during their pregnancy, if they were treated with beta blockade prior to pregnancy and discontinued therapy, or if the patients were receiving beta blockers but it was not reported in the literature.

However, it is notable that these patients presented with an aortic dissection during pregnancy while being managed with beta blockers. This questions the efficacy of beta blockers to prevent AAD during pregnancy in women with MFS. Several studies describe cases of MFS women who received beta-blocker therapy during their pregnancy without adverse cardiovascular outcomes. Omnes et al. describes a prospective study of pregnancies in a small group of MFS patients ($n = 18$). Fifteen of these patients were treated with beta blockade. Notably, 1 woman had an increase in aortic root diameter of >10% during her pregnancy despite beta blockade. The only aortic dissection in this cohort was seen in a patient that was not treated with beta blockers.[26] However, the sample size of this study was small and does not convincingly indicate that beta blockers are efficacious for the prevention of aortic dissection in pregnant Marfan patients.

The relative lack of information regarding beta blockade in women with MFS who developed aortic dissections during pregnancy presents a clinical challenge that should be investigated further. The potential for negative effects on the fetus highlights the importance of maintaining a balance between safeguarding the mother's prognosis and the avoidance of fetal growth restriction. We understand that some women may want to avoid the use of any medication during their pregnancy, especially if there is not strong evidence to support the use of this medication. For this reason, we believe that the decision to continue beta-blocker therapy during pregnancy should be a shared decision between the physician and the patient, considering factors such as aortic root diameter, aortic root growth rate, risk for intrauterine growth restriction, and patient concerns.

4.4 | Limitations

We understand that this analysis, particularly its format as a literature review, presents several limitations. First, we are limited to case reports published in the literature. We understand that a publication bias may exist, with negative outcomes being less likely to be published. Additionally, there may be a predominance of interesting or unique cases published in the literature. We understand that these cases may not adequately represent the population or the current clinical environment surrounding AAD in pregnancy. As such, the analysis in this review is meant to address only the current literature.

Another consideration to note is that for many of the cases, aortic root diameter and beta blocker usage were not described. It is unclear if aortic root measurements were not obtained or if they were simply not included in the report. Similarly, it is unclear from the literature if the MFS patients were being treated with beta blockers and the information was omitted or if the patients were not taking beta blockers.

An additional limitation of this study is the obtainment of postpartum cases. We defined a postpartum event as a dissection occurring 1 year after delivery. As such, it is possible that some case reports within the 12-month timeline may not have been cross-referenced with "pregnancy" and were missed by our search methods.

4.5 | Future directions

The most significant development on this topic would be the identification of women who are at risk for AAD during pregnancy. In the case of Marfan syndrome, this could be accomplished by earlier diagnosis of MFS. Additional analysis of the specific Ghent criteria met by Marfan women who have pregnancy-related dissections may illuminate key features that this population shares. If these women share a specific phenotype, then clinicians may be better equipped identify Marfan patients at high risk for pregnancy-related dissections. Several articles depicted cases of pregnancy-related AAD in women with undiagnosed MFS. Three of these cases described women who did not meet any of the Ghent criteria other than the AAD and a positive genetic test. For these patients, the development of additional Ghent criteria (if possible) may be helpful for earlier diagnosis of MFS. If additional Ghent features can be identified for these women, then obstetricians could implement Marfan screening tests for their patients during their first encounter.

Current guidelines focus on aortic root diameter as the strongest risk factor for the development of pregnancy-related AAD in women with MFS. In our analysis, we found a surprising number of cases of AAD in pregnant Marfan women with aortic root diameter <45 mm. These cases question the current understanding that the risk of AAD during pregnancy in Marfan patients is a direct function of aortic root diameter. Further evaluation is needed to identify the risk factors that contribute to AAD during pregnancy as well as the pathophysiology behind these events. One interesting proposal is that some women

with Marfan syndrome have an accelerated growth of the aortic root during pregnancy. Additional studies with serial aortic root measurements throughout pregnancy and the postpartum period may identify factors that lead to this type of aortic root growth in Marfan women.

This review focuses primarily on pregnancy-related AAD in women with MFS, but we do not want to underscore the importance of identifying non-Marfan women who are at risk for this event. There is a paucity of studies that evaluate the factors leading to AAD during pregnancy in women without underlying connective tissue disease. Future studies addressing these factors may allow obstetricians to identify which patients may be at risk for dissection.

Finally, the importance of beta blockers in the management of MFS has been a topic of debate. There remains very little evidence supporting the idea that beta blockers are efficacious for attenuating aortic root growth and preventing aortic dissection during pregnancy in MFS. Because beta blockers carry a risk to the fetus (FDA Pregnancy Category C), we believe that there is a need for additional studies regarding the use of these medications in pregnant Marfan women.

CONFLICT OF INTEREST

The authors report no conflicts of interest.

AUTHOR CONTRIBUTIONS

Concept/design: Smith
Data analysis/interpretation: Smith
Drafting article: Smith
Concept/design: Gros
Critical revision of article: Gros

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