Endovascular thoracic aortic repair in confirmed or suspected genetically triggered thoracic aortic dissection

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ABSTRACT

Objective: Endovascular repair in patients with connective tissues disorders is not recommended because of concern for repair failure. The aim of this study was to investigate thoracic endovascular aortic repair (TEVAR) outcomes in patients with confirmed or suspected syndromic and nonsyndromic genetically triggered thoracic aortic dissection.

Methods: We analyzed data for patients with descending thoracic aorta (DTA) dissection treated with TEVAR from the National Registry of Genetically Triggered Thoracic Aortic Aneurysms and Cardiovascular Conditions (GenTAC). Enrolled patients had confirmed (syndromic or familial) or suspected genetically triggered thoracic aortic disease. The latter group includes patients with sporadic aortic dissection presenting at 50 years of age or younger in the absence of a family history or syndromic features.

Results: Between 2006 and 2014, there were 371 patients with DTA dissection enrolled in GenTAC. TEVAR was performed in 31 cases (58.1% male; median age, 47 years; range, 21.3-65.6 years). Genetically triggered aortic dissection was confirmed in 18 cases, and an additional 13 cases had suspected genetically triggered dissection because of early onset of presentation. TEVAR was performed in nine patients with type A aortic dissection: five in conjunction with acute type A dissection repair and four in the chronic phase to treat aneurysmal degeneration of the residual dissected DTA (median interval to TEVAR, 2.1 years). TEVAR was also performed in 22 cases of type B aortic dissection (TBAD), 12 acute and 10 chronic (median interval to TEVAR, 1.6 years). There were no perioperative deaths. Median follow-up for all cases was 2 years (range, 0.4 month-7 years). Reinterventions after TEVAR were performed in 13 cases (41.9%). This included urgent repair of three retrograde ascending aorta dissections occurring after TEVAR for acute TBAD (25%) and seven thoracoabdominal repairs with stent graft explantation (22.6%) at a median of 7 months after TEVAR (range, 1-16.6 months).

Conclusions: TEVAR in patients with genetically triggered aortic dissections can be lifesaving in the acute setting though associated with high risk of retrograde aortic dissection in acute TBAD. For chronic dissection-related DTA aneurysmal degeneration, TEVAR could potentially be lifesaving in patients deemed too high risk for open surgical repair. Close postoperative surveillance is required, given the risk of subsequent device failure and need for reintervention. Because these circumstances are rare, multicenter prospective enrollment of patients with genetically triggered aortic disease is essential to delineate the indications for and risks of TEVAR in this heterogeneous population. (J Vasc Surg 2018;68:364-71.)

The use of thoracic endovascular aortic repair (TEVAR) in treating acute type B aortic dissection (TBAD) was first reported in 1999 and has since become a game changer in the management of acute TBAD.[1] TEVAR is the treatment of choice in complicated acute TBAD.[2] Few data are available to guide treatment in patients with genetically triggered aortic dissections. The consensus statement of the Society of Thoracic Surgeons

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recommends strongly against endovascular repair in patients with connective tissue disorders unless operative risk is deemed truly prohibitive by a center experienced in management of complex aortic disease and in cases of aortic rupture.[3] The last European guidelines on diagnosis and treatment of aortic diseases recommend open surgical repair over TEVAR in patients with Marfan syndrome (MFS) except as an emergency bridge to definitive surgical repair.[4] These recommendations stem from concerns for device fixation failure in a diseased aorta. The chronic outward radial force exerted by the stent graft on the diseased aorta leads to future aortic dilation, loss of seal, development of endoleaks, and device migration or erosion. In addition, stent graft-induced new entry tears and retrograde ascending aorta dissection have been reported in association with TEVAR. These modes of device failure lead to subsequent need for reoperation, stent graft explantation, and open repair of the aorta.[5-7]

What is less clear is the role of TEVAR in genetically triggered aortic disease due to gene mutations other than those in MFS. In addition, patients with aortic dissection at a young age are suspected of having genetically triggered aortic disease, and similarly, the role of TEVAR remains unclear in this population.[8]

The aim of this study was to investigate the outcomes of TEVAR in patients with confirmed or suspected genetically triggered thoracic aortic dissection enrolled in the National Registry of Genetically Triggered Thoracic Aortic Aneurysms and Cardiovascular Conditions (GenTAC).[8]

METHODS

GenTAC[8] is a multi-institutional National Institutes of Health-funded registry funded from 2006 to 2015. The registry enrolled patients from eight regional clinical centers with a confirmed or suspected genetically triggered thoracic aortic aneurysm or dissection.[6] The participating centers included Baylor College of Medicine, Johns Hopkins University School of Medicine, National Institute on Aging at Harbor Hospital, Oregon Health & Science University, Queens Medical Center, University of Pennsylvania Health System, Weill Medical College of Cornell University, and University of Texas Health Science Center at Houston McGovern Medical School. Institutional Review Board approval was obtained at each of the participating centers, and individual informed consent was obtained from each enrolled patient. The registry design included retrospective data abstraction at the time of enrollment with prospective observational follow-up data. Data were abstracted from the medical charts and patient questionnaires.

The registry was queried for the diagnosis of "ever dissected descending thoracic aorta" and "thoracic endovascular repair." The deidentified data were then requested for analysis.

ARTICLE HIGHLIGHTS

- **Type of Research:** Retrospective analysis of prospectively collected National Registry of Genetically Triggered Thoracic Aortic Aneurysms and Cardiovascular Conditions (GenTAC) data
- **Take Home Message:** In 31 patients with descending thoracic aorta dissection treated with thoracic endovascular aortic repair, 13 (42%) required reintervention at a median of 7 months, 3 for retrograde ascending aorta dissection and 7 for thoracoabdominal aortic aneurysm repairs with stent graft explantation.
- **Recommendation:** This study suggests that patients with genetically triggered descending thoracic aorta dissection, treated with thoracic endovascular aortic repair, have a high risk of retrograde aortic dissection and reintervention.

Inclusion criteria included patients with confirmed or suspected genetically triggered aortic disease, defined as follows:

- Syndromic aortic dissection: patients with gene mutations leading to syndromic disorders such as MFS, Loeys-Dietz syndrome, and vascular Ehlers-Danlos syndrome.
- Nonsyndromic genetically triggered aortic dissection: patients with gene mutations leading to nonsyndromic heritable thoracic aortic disease, such as mutations in the *FBN1*, *TGFBR1*, *TGFBR2*, *ACTA2*, and *MYH11* genes.[8]
- Familial dissections: patients with a first-degree (parent, child, sibling) or second-degree (cousin, aunt, uncle, grandparent) relative with an aortic aneurysm or dissection. These patients can have syndromic aortic disease (MFS, Loeys-Dietz syndrome, vascular Ehlers-Danlos syndrome) or nonsyndromic genetically triggered aortic disease. Moreover, these patients could have one of the known mutations leading to heritable thoracic aortic disease or a mutation that has yet to be discovered.
- Early-onset dissection: patients with sporadic aortic dissection presenting at an early age (50 years or younger) in the absence of a family history or syndromic features. These patients are *suspected* of having an underlying genetic etiology.

Exclusion criteria for GenTAC included the inability of the patient to provide consent for enrollment in the study.[8]

Analysis. Statistical analysis was performed using SPSS 19.0 for Windows (IBM Corp, Armonk, NY). Continuous data are presented as means and standard deviations or medians and ranges, as appropriate. Means of continuous data were compared using the Student *t*-test. Categorical variables were compared using the χ^2 and

Table I. Reasons for enrollment of 31 patients with descending thoracic aorta (DTA) dissection treated with thoracic endovascular aortic repair (TEVAR) in the National Registry of Genetically Triggered Thoracic Aortic Aneurysms and Cardiovascular Conditions (GenTAC)

	No. (%)
Genetically triggered dissection	
Syndromic	
MFS	7 (22.6)
Loeys-Dietz syndrome	1 (3.2)
Vascular Ehlers-Danlos syndrome	1 (3.2)
Nonsyndromic	
ACTA2 mutation	1 (3.2)
Family history[a]	8 (25.8)
Suspected to be genetic	
Early dissection onset[b]	13 (41.9)

MFS, Marfan syndrome.
[a]Patients with familial aortic disease who are nonsyndromic. These patients have a first-degree (parent, child, sibling) or second-degree (cousin, aunt, uncle, grandparent) relative with a thoracic aortic aneurysm or dissection without syndromic features.
[b]Sporadic aortic dissection at age 50 years or younger in the absence of a family history or syndromic features.

Fisher exact tests as appropriate. All statistical tests were two sided, and a *P* value $<.05$ was considered statistically significant. Deidentified data and analyses were performed and stored in a password-protected workspace.

RESULTS

Between 2006 and 2014, there were 3699 patients enrolled in GenTAC. Descending thoracic aorta (DTA) dissection was noted in 371 patients (65.2% male). These included 163 cases of type A aortic dissection and 208 cases of TBAD. TEVAR was performed in 31 (8.4%) cases (58.1% male; median age, 47 years; range, 21.3-65.6 years). The indication for TEVAR was to repair the dissected DTA due to type A aortic dissection (DeBakey I) in nine cases and TBAD in 22 cases. Patients had genetically triggered dissection in 18 cases, 10 with a syndromic aortic dissection and 8 with familial dissections without a known mutation. An additional 13 patients had sporadic dissections with a suspected genetic component because of early dissection onset (at age 50 years or younger) in the absence of a family history or syndromic features (Table I).

The majority of TEVAR cases (n = 24 [77.4%]) were performed at a median of 7.3 months before enrollment in GenTAC. Three cases were identified and enrolled at the time of TEVAR (9.6%), and five cases had TEVAR performed after GenTAC enrollment (16.2%). The median follow-up after TEVAR was 2 years (range, 0.4 month-7 years). Table II summarizes the demographics and comorbid conditions of the cohort and includes a comparison of patients with genetically triggered aortic dissection and early dissection onset. Patients with genetically triggered dissection were slightly younger than patients with early dissection onset (not statistically significant) and had a lower percentage of African American and Asian patients ($P = .027$).

TEVAR in cases of type A aortic dissection. TEVAR was performed to repair the DTA dissection in nine patients with type A aortic dissection (mean age, 43 ± 10 years; 55.6% male). Among those, the dissection was genetically triggered in eight cases and suspected to be of genetic etiology (early dissection onset) in one case. The timing of TEVAR varied. In five cases, TEVAR was performed in conjunction with repair of acute type A aortic dissection (including arch repairs: hemiarch repair in four cases and full arch repair in one case). In four cases, TEVAR was used to treat chronic DTA aneurysmal degeneration associated with the type A aortic dissection at a median interval of 2.1 years (range, 5.5 months-13 years) after acute type A dissection repair (including arch repairs: hemiarch repair in 3 cases and full arch repair in 1 case). Data on size of DTA were available for three cases; the DTA diameter at TEVAR measured 4.1 cm, 6 cm, and 8 cm. Median follow-up after TEVAR was 11 months (range, 0-4 years). Three cases required stent graft explantation and open thoracoabdominal aortic aneurysm repair (33.3%) as detailed in Table III.

TEVAR in cases of TBAD. TEVAR was performed in 22 cases of TBAD, 10 with genetically triggered dissection and 12 with early-onset dissection. The repair was performed in the acute phase for 12 cases and in the chronic phase to treat DTA aneurysmal degeneration in 10 cases. The demographics, comorbid conditions, and repair details are summarized in Table IV. The median interval to repair in chronic TBAD was 1.6 years (range, 5 weeks-28 years) after TBAD. In two cases, a previous DTA repair preceded TEVAR—a patient with MFS (prior hemiarch and DTA repair) and a patient with familial dissection (prior DTA repair). None of the cases had prior elephant trunk repairs. The median DTA diameter at chronic TBAD repair was 4.2 cm (range, 3-8 cm, eight cases only). There were no perioperative deaths. Postoperative complications included prolonged intubation (n = 3), acute renal failure (n = 1), and retrograde dissection extending into the ascending aorta (n = 3). All retrograde dissection cases occurred after TEVAR for acute TBAD (25%) and are detailed as follows:

- A 46-year-old man presented with an acute TBAD in a 2.9-cm-diameter DTA and an arch entry tear. He developed a retrograde aortic dissection after TEVAR into an aneurysmal ascending aorta and root (unknown diameter). This was treated with a root, ascending, and hemiarch repair.
- A 47-year-old woman presented with acute TBAD in a 5.9-cm-diameter DTA. The retrograde dissection was treated with ascending thoracic aortic and full arch repair.

Table II. Demographics and comorbid conditions of 31 patients enrolled in the National Registry of Genetically Triggered Thoracic Aortic Aneurysms and Cardiovascular Conditions (*GenTAC*) with descending thoracic aorta (DTA) dissection treated with thoracic endovascular aortic repair (*TEVAR*)

	Cohort (N = 31)	Genetically triggered dissection (n = 18)	Early dissection onset[a] (n = 13)	*P*
Age at dissection, years	42.9 (±11.2)	42 (±12.6)	46.6 (±9.1)	.087
Age at GenTAC enrollment, years	46.4 (±11.2)	46.3 (±12)	48.4 (±9.9)	.271
Age at TEVAR, years	45.6 (±11)	45.7 (±11.9)	47.3 (±9.9)	.258
Male	18 (58.1)	9 (50)	9 (69.2)	.289
Race				.027
White, non-Hispanic	21 (67.7)	16 (88.9)	5 (38.5)	
White, Hispanic	1 (3.2)	0	1 (7.7)	
African American	6 (19.4)	1 (5.6)	5 (38.5)	
Asian	3 (9.7)	1 (5.6)	2 (15.4)	
Comorbid conditions				
Hypertension	23 (74.2)	12 (66.7)	11 (84.6)	.260
Coronary artery disease	3 (9.7)	1 (5.6)	2 (15.4)	.361
Valve dysfunction	14 (45.2)	11 (61.1)	3 (23.1)	.036
Current smoker	0	0	0	
Smoked at least 100 cigarettes	9 (29)	9 (50)	0	.002
Family history	13 (41.9)	13 (72.2)	0	N/A
Dissection type				.026
Type A aortic dissection	9 (29)	8 (44.4)	1 (7.7)	
TBAD	22 (71)	10 (55.6)	12 (92.3)	

N/A, Not applicable; *TBAD*, type B aortic dissection.
Categorical variables are presented as number (%). Continuous variables are presented as mean ± standard deviation.
[a]Age 50 years or younger in the absence of a family history or syndromic features.

Table III. Reinterventions after thoracic endovascular aortic repair (TEVAR) for descending thoracic aorta (*DTA*) repair in patients with genetically triggered type A aortic dissection

Age, years; sex; diagnosis	Interval to reintervention	DTA diameter, cm	Reintervention details
46.3; female; vascular Ehlers-Danlos syndrome In conjunction with acute type A dissection repair	321 days	NR	Stent graft explantation, open TAAA II repair
40.3; male; Loeys-Dietz syndrome Type A, chronic	16.6 months	8	Stent graft explantation and open TAAA I repair for false lumen expansion
27.5; male; MFS Type A, chronic	2.1 months	4.1	Stent graft explantation and open TAAA II repair for type Ia and type Ib endoleak, direct communication between stent graft and false lumen

MFS, Marfan syndrome; *NR*, not reported; *TAAA*, thoracoabdominal aortic aneurysm.

- A 51-year-old man with an *ACTA2* mutation presented with a ruptured TBAD in a 3.1-cm-diameter DTA. He was treated with zone 2 coverage and subsequently developed retrograde aortic dissection (Fig) with cerebral and peripheral malperfusion requiring emergent ascending and arch repair.

Follow-up information was available for 21 cases with a median follow-up of 2.3 years (range, 1 month-7 years). Reintervention procedures were performed in 10 cases (45.5%) at a median of 1.3 months after TEVAR (range, 0.2-16.5 months) as summarized in Table V. Six of the reinterventions were undertaken in patients treated with TEVAR for acute TBAD (50% of the acute TBAD cases), including the three cases with retrograde dissection detailed earlier. There were two deaths in this cohort unrelated to the TEVAR or revision procedures. One patient with a familial aortic dissection died at the age of 47.6 years of "atherosclerotic cardiovascular disease."

Table IV. Demographics, comorbid conditions, and repair details of 22 patients with confirmed or suspected genetically triggered type B aortic dissection (*TBAD*) treated with thoracic endovascular aortic repair (*TEVAR*)

	Genetically triggered dissection (n = 10)	Early dissection onset (n = 12)	*P*
Male	5 (50)	8 (66.7)	.429
Race			.043
White, non-Hispanic	9 (90)	4 (33.3)	
White, Hispanic	0	1 (8.3)	
African American	0	5 (41.7)	
Asian	1 (10)	2 (16.7)	
Comorbid conditions			
Hypertension	7 (70)	10 (83.3)	.457
Coronary artery disease	1 (10)	2 (16.7)	.650
Valve dysfunction	5 (50)	3 (25)	.225
Smoked at least 100 cigarettes	3 (30)	0	.041
Prior type A dissection (DeBakey II)	3 (30)	2 (16.7)	
Family history	6 (60)	0	—
Age at dissection, years	41.5 (±12.5)	46.3 (±9.4)	.315
Age at TEVAR, years	46.3 (±13.2)	48.2 (±10.3)	.903
Follow-up duration, months	33 (±27)	28.8 (±24.7)	.707
Acute TBAD[a]	4	8	
Prior root or ascending aortic repair	1 (25)	2 (25)	—
Prior type A aortic dissection (DeBakey II)	1 (25)	1 (12.5)	
Carotid-subclavian bypass	0	1 (12.5)	—
Retrograde dissection	1 (25)	2 (25)	—
Reintervention	3 (75)	3 (37.5)	—
Type I endoleak	0	1 (12.5)	—
Stent graft explantation	0	2 (25)	—
Chronic TBAD[a]	6	4	
Maximum DTA diameter, cm	4.4 (±1.6)	5.3 (±2.3)	—
Prior root or ascending aortic repair	4 (66.7)	2 (50)	—
Prior arch repair	1 (16.7)	0	
Prior DTA repair	2 (33.3)	0	—
Carotid-subclavian bypass	0	2 (50)	—
Reintervention	2 (33.3)	2 (50)	—
Type I endoleak	1 (16.7)	2 (25)	—
Stent graft explantation	1 (16.7)	1 (25)	—

DTA, Descending thoracic aorta.
Categorical variables are presented as number (%). Continuous variables are presented as mean ± standard deviation.
[a]Comparisons were not made because of the small number of cases.

The second was a patient with an early-onset TBAD who died of unknown cause 5 years after TEVAR at the age of 55 years.

DISCUSSION

We reviewed TEVAR outcomes in a cohort of patients with confirmed (syndromic or familial) or suspected (early onset) genetically triggered aortic dissections. This study highlights the current concern regarding retrograde aortic dissection when TEVAR is used to treat acute TBAD in this population of patients. Whereas TEVAR may be lifesaving in this circumstance, it is associated with a high risk (25%) of retrograde aortic dissections in patients with confirmed or suspected genetically triggered dissection when the stent graft is placed in an unrepaired arch. Retrograde aortic dissection is a well-known complication after TEVAR for acute TBAD, with a relatively rare incidence ranging between 1.3% and 4%.[9-12] Several factors have been implicated in this complication, including stent graft oversizing, failure

Table V. Reintervention performed after thoracic endovascular aortic repair (*TEVAR*) in patients with confirmed or suspected genetically triggered type B aortic dissection (TBAD)

Type	Age, years; sex; diagnosis	Interval to reintervention	Maximum DTA diameter, cm	Reintervention details
Acute	29.7; female; familial	30 days	6.4	Stent graft explantation and open TAAA I repair for type II endoleak
	52.6; male; MFS	NR	NR	Stent graft explantation for stent graft collapse
	51; male; *ACTA2* mutation	6 days	3.1	Root and ascending aortic repair for retrograde type A dissection (Fig)
	45.5; male; early onset	42 days	2.9	Root and ascending aortic repair for retrograde type A dissection
	46.9; female; early onset	1 day	5.9	Coil embolization of false lumen type Ia and type Ib endoleaks; retrograde dissection
	51.9; male; early onset	0	NR	External iliac artery stent for renal and lower extremity malperfusion
Chronic	21.3; female; MFS	0.4 month	2.8	Second TEVAR placed inside the first
	46; male; familial	16.5 months	6.8	Stent graft explantation and open TAAA I repair for type Ib endoleak
	25.3; female; early onset	4.0 months	6.2	Second TEVAR, then stent graft explantation and open TAAA I repair for type Ia endoleak at the arch and aneurysmal degeneration to 7.9 cm
	50.8; male; early onset	0.2 month	3.9	Coil embolization for type II endoleak at left subclavian artery

DTA, Descending thoracic aorta; *MFS*, Marfan syndrome; *NR*, not reported; *TAAA*, thoracoabdominal aortic aneurysm.

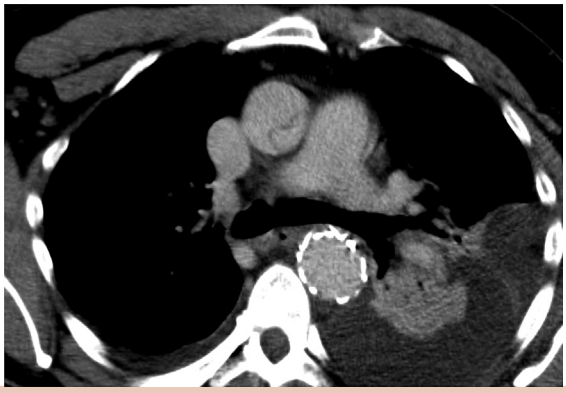

Fig. Retrograde dissection in the ascending thoracic aorta of a 51-year-old man with *ACTA2* mutation treated with zone 2 coverage thoracic endovascular aortic repair (TEVAR) for a ruptured type B aortic dissection (TBAD).

to cover the tear site during stent graft treatment, ballooning of the dissected aorta, and fragility of the aortic wall. The last concern specifically applies to patients with genetically triggered aortic dissections.

Our study also demonstrates the high rates of subsequent secondary interventions after TEVAR, including stent graft explantations and open aortic repair. These rates are in line with previously reported findings in MFS cases. In a series of 69 MFS patients, 20.3% of MFS patients treated with TEVAR for TBAD required secondary endovascular interventions and 21.7% underwent subsequent open repair of the aorta compared with 12% to 14% reintervention rates after TEVAR for sporadic TBAD.[5-7,12-17]

These findings highlight the inadequacies in current stent graft design to treat patients with genetically triggered aortic disease. Open repair remains the "gold standard" in patients with confirmed or suspected dissections who carry an acceptable operative risk and who have not had multiple aortic operations. In contrast, TEVAR is perhaps better suited for treating specific problems in this population of patients, such as exclusion of focal pseudoaneurysms when an existing aortic graft serves as the proximal and distal landing zones, fixation of TEVAR in an elephant trunk graft after previous arch repair, or extension of a device distally from a secure thoracic replacement. It can also be an option for patients with prohibitive operative risk that precludes an open repair or a temporizing lifesaving intervention in cases of aortic rupture. The high percentage of cases requiring secondary interventions among patients with confirmed and suspected genetically triggered dissection, including explantations and open repair, highlights the importance of perioperative counseling to emphasize that these repairs are often a temporizing measure and require lifelong follow-up with surveillance imaging.[3,17-20]

There are several limitations to this study inherent to design of the registry. First, a small group of patients with heterogeneous aortic diseases and chronicity was reviewed (type A and B aortic dissection, acute and chronic phase repairs). Second, patients were enrolled in the registry on the basis of their genetic diagnosis

rather than by the surgical intervention. This led to enrollment at different time points in the disease process; some patients were enrolled at the time of the acute dissection, whereas others were enrolled at the time of aortic repair (TEVAR or open repair). This design limitation leads to a wide range of follow-up durations, missing operative details, and a possibility of missing some patients who would have met inclusion criteria. With the exception of patients enrolled by age (early dissection onset), the nature of GenTAC enrollment precludes ascertainment and enrollment of patients who died early of dissection or surgical complications before establishment of a genetic diagnosis. Third, whereas data were abstracted from the medical records and patient questionnaires, the operative details of TEVAR, such as type of graft, degree of oversizing, size of the aorta at the time of repair, and ballooning after stent graft deployment, were not recorded consistently, thus limiting our ability to further comment on the technical details. In addition, the size of the aorta at the time of dissection was not recorded consistently, and we were unable to ascertain the exact reasons for the secondary interventions beyond what has been presented. Last, imaging was not available for the majority of the patients; thus, we were unable to ascertain the frequency of imaging follow-up and, in the cases of endoleaks, the exact nature of the endoleaks (eg, true type Ib endoleak vs persistent perfusion of the distal false lumen) beyond what was abstracted from the medical records.

Despite these limitations, the study provides valuable information. The data demonstrate that TEVAR can be lifesaving in the acute phase of TBAD. In addition, although one cannot make broad generalizations based on limited data, these data offer an opportunity to reflect on using stent grafts in patients with sporadic aortic dissections occurring at a young age, suggesting a genetically triggered aortic dissection. This is an opportunity to discuss the best approach for each individual patient (open repair vs TEVAR with close follow-up). Moreover, the lessons learned from this study design inform future prospective observational multicenter studies, such as the Montalcino Aortic Consortium.[21] Multicenter prospective enrollment of patients with genetically triggered DTA dissection is essential to delineating the natural history of aortic dissection according to the underlying mutation, which is a knowledge gap in our contemporary literature. This information is essential to accurately phenotype the increasing number of identified heritable thoracic aortic disease mutations and will be the basis for providing clinical management recommendations and device design considerations.

CONCLUSIONS

TEVAR in patients with genetically triggered aortic dissections can be lifesaving in the acute setting though associated with high risk of retrograde ascending aortic dissection in acute TBAD. In cases of dissection-related DTA aneurysmal degeneration, TEVAR could potentially be lifesaving in patients deemed too high risk for open surgical repair. Close postoperative surveillance is required, given the risk of subsequent device failure and need for reintervention. Because these circumstances are rare, multicenter prospective enrollment of patients with genetically triggered aortic disease is essential to delineate the indications for and risks of TEVAR in this heterogeneous population.

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AUTHOR CONTRIBUTIONS

Conception and design: SS, SL, DM
Analysis and interpretation: SS, KE, SL
Data collection: KE, FA, SL
Writing the article: SS, KE
Critical revision of the article: SS, KE, FA, SL, DM
Final approval of the article: SS, KE, FA, SL, DM
Statistical analysis: SS
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Overall responsibility: SS

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