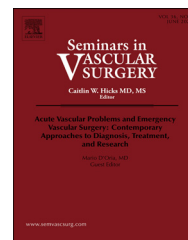


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Review article

Emergency vascular surgical care in populations with unique physiologic characteristics: Pediatric, pregnant, and frail populations



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ABSTRACT

Vascular surgical emergencies are common in vascular surgical care and require complex decision making and multidisciplinary care. They are especially challenging when they occur in patients with unique physiological characteristics, such as pediatric, pregnant, and frail patients. Among the pediatric and pregnant population, vascular emergencies are rare. This rarity challenges accurate and timely diagnosis of the vascular emergency. This landscape review summarizes these three unique populations' epidemiology and emergency vascular considerations. Understanding the epidemiology is the foundation for accurate diagnosis and subsequent management. Considering each population's unique characteristics is crucial to the emergent vascular surgical interventions decision making. Collaborative and multidisciplinary care is vital in gaining expertise in managing these special populations and achieving optimal patient outcomes.

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1. Introduction

Emergency vascular care is a common aspect of a vascular surgeon's practice, and clinical decision making is often complex and requires multidisciplinary care. Complexity increases when caring for patients with unique physiologic

characteristics, such as pediatric, pregnant, and frail individuals. In this landscape review, we summarize the epidemiology and considerations for emergency vascular management of these patient groups. Understanding the unique epidemiology is the foundation for accurate diagnosis and management in these populations. When making decisions about emergent vascular surgical interventions, it is also crucial to consider the unique characteristics of each population. In the case of pediatric patients, we will discuss traumatic vascular injuries, catheter-related acute limb ischemia (ALI), and aortic

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dissections (AD). For pregnant patients, we will focus on traumatic vascular injuries, ALI, AD, splenic artery aneurysms (SAAs), and thromboembolic complications. Finally, for frail patients, we will highlight the assessment of frailty and the role of palliative care in the vascular emergency setting.

2. Emergency vascular surgical care in the pediatric population

Vascular emergencies are rare in the pediatric population, and pediatric vascular operations are generally uncommon. Emergency vascular surgical care is required in cases of trauma, ALI, and aortic or arterial aneurysms and dissections among infants and children. As many free-standing children's hospitals lack adult vascular surgery expertise, the care of vascular surgery injuries in the pediatric population is surgically managed by pediatric, vascular, trauma, and orthopedic surgeons. Given the rarity of vascular surgical needs, there are no specific training competencies for pediatric vascular surgical care in vascular or pediatric surgery specialty training [1–3]. Data are sparse regarding vascular surgery practice patterns by specialty. A single-center retrospective review of 94 pediatric trauma patients (65.9% were male patients) between 1993 and 2015 demonstrated changing patterns of specialty care provided, with vascular surgeons performing none of the surgical repairs in 1993–2004 to 23% of the cases in 2005–2015 [3].

2.1. Pediatric vascular trauma

2.1.1. Pediatric vascular trauma epidemiology

Unintentional injury is the leading cause of death among children and adolescents aged 1 through 16 years [4]. The Centers for Disease Control and Prevention reported a 2001–2020 crude rate of 6.9 deaths per 100,000 children in this age group [4]. Traumatic vascular injuries are rare among the pediatric population, with an incidence of 0.6% of pediatric trauma, less than what is seen in the adult population (1.6%; $P < .001$) [5]. A similar finding was observed in a 2021 single-center experience reported by Perea et al [6] found that vascular injuries accounted for 0.8% of all pediatric trauma. Among 22,089 vascular injuries in the American College of Surgeons National Trauma Data Bank (NTDB) from 2002 to 2006, 1,187 were among individuals younger than 16 years, thus accounting for 5.4% of all vascular trauma [5].

Most of the injured patients were male (73.7%) [7]. Blunt trauma was the predominant mechanism of injury, accounting for 57% of the cases in NTDB [5,7]. In a single-center study of 60 pediatric trauma patients, 78.3% of the patients were male and vascular injuries due to blunt trauma accounted for 70% of the cases among patients younger than 14 years [6]. Penetrating trauma was more common among pediatric patients aged 14 through 18 years at the same center, accounting for 67.5% of the pediatric trauma cases [6]. In this cohort, gunshot wounds were the most common mechanism of vascular injury, affecting 36.7% of the injured pediatric patients younger than 18 years [6]. The leading cause of death was firearm injuries, followed by motor vehicle crashes (MVCs), noted at 36.9% and 34.6%, respectively, in the NTDB data

[7]. Similar findings were also reported in single-institutional studies [3,6,8].

Upper extremity vessels were the most commonly injured (approximately 34.9%) [5,7]. The least commonly injured vessels were the thoracic vessels (7.5%) [5,7].

The California Office of Statewide Health Planning and Development patient discharge database for 2007–2014 showed that of 577 upper extremity arterial injuries among patients younger than 18 years (78.3% were male), the brachial artery was the most commonly injured (31.7%) and the subclavian artery was the least commonly injured (4.6%) [9]. In-hospital mortality was highest among pediatric patients; injured subclavian and axillary arteries were at 11.1% and 6.3%, respectively [9]. None of the patients with upper extremity arterial injuries had a major amputation [9]. An injury notable in children was the supracondylar humeral fracture. This injury is associated with an 8% to 12% risk of brachial artery injury [10]. The risk of brachial artery injury is increased when there is a posterolateral displacement of the humerus fracture [11]. Examination findings consistent with the arterial injury include a cool pale pulseless hand. The presence of antecubital fossa ecchymosis is suggestive of brachial artery injury [12]. The injury can be associated with median and anterior interosseous nerve injury, and long-term complications include Volkmann's ischemic contracture risk [10–12].

Lower extremity vessel injury affects an estimated 21% of pediatric trauma cases and carries higher mortality and amputation rates than upper extremity vessel injury [7,9,13]. The California Office of Statewide Health Planning and Development patient discharge database for 2007–2014 showed that of 274 lower extremity arterial injuries among patients younger than 18 years (82.1% were male), the superficial femoral artery was the most commonly injured (39.1%) [9]. The anterior tibial artery was the least commonly injured (10.9%) [9]. The study also demonstrated variation in risk of in-hospital mortality ranging from none, in cases of tibial arterial injuries, to 8.8% in-hospital mortality among patients with injured common femoral arteries. Major amputation also varied by the location of the injury, with the highest amputation rates associated with anterior tibial artery injuries (6.7%), followed by popliteal artery injuries (4.5%) and superficial femoral artery injuries (3.7%) [9].

Abdominal vascular injuries are the leading cause of death in the pediatric trauma population (42.1% of the cases in the NTDB data) [7]. Abdominal vessel injuries account for 20.1% of the cases, affecting the inferior vena cava, iliac (Fig. 1), and renal vessels most frequently [5,7]. Abdominal aortic injuries, especially blunt abdominal aortic injuries (BAAIs), are rare, not unlike the adult population [14]. BAAI commonly results from an MVC, often present with a seat belt sign, and are associated with hollow viscus perforation and spine fractures [15]. Less frequently, they can also present with paraplegia, as detailed in Table 1, which summarizes the presentation and management of pediatric BAAI case reports [15–36]. Unfortunately, the long-term outcomes reported in this population are limited.

2.1.2. Pediatric vascular trauma management

Pediatric vascular trauma management principles include early identification and control of exsanguinating hemorrhage, especially in chest and abdomen injuries due to high-

Table 1 – Presentation and management of pediatric blunt abdominal aortic injury case reports.

First author	Year	Age (y)/sex	Injury description	Management and outcome
Kory [16]	2000	2.5/M	Aortic thrombosis, paraplegia	Surgical exploration, thrombectomy, residual paraplegia
Inaba [17]	2001	8/M	2-cm infrarenal abdominal aorta dissection, leg weakness	Non-operative, mild left weakness at 6 mo
Lin [18]	2003	6/F	Distal aortic transection, leg weakness	Hypogastric artery patch repair, neurologic recovery at 3 mo
Milas [19]	2003	6/F	Partial transection and thrombosis, paraplegia	Thrombectomy and intimal repair, residual paraplegia at 21 d
Soares [20]	2003	6/F	Infrarenal abdominal aortic transection, normal motor examination	8-mm interposition Gore-Tex (W. L. Gore and Associates) graft
Muniz [21]	2004	8/M	Infrarenal abdominal aortic transection, normal motor examination	Postoperative paraplegia, 6-mo follow-up
Prince [22]	2004	9/F	Distal aortic dissection, paraplegia	12-mm bifurcated Hemashield aortic bi-iliac graft, no follow-up
Milas [19]	2004	11/NR	Distal aortic “disruption”	PTFE interposition graft, recovery, 21-mo follow-up
Diaz [23]	2006	3/M	Delayed presentation 10 y later with aortic stenosis due to web formation	Web excision and bovine pericardium patch angioplasty, recovery, 4-mo follow-up
Choit [15]	2006	9/F	Distal aortic transection	16-mm Hemashield interposition graft, recovery, 4 y follow-up
		11/M	Distal aortic pseudoaneurysm	Non-operative, stable with 2 y of follow-up
		12/F	Aortic bifurcation transection, paraplegia	Bifurcated PTFE aortic bi-iliac graft, 18 d follow-up
Aidinian [24]	2006	10/M	Aortic transection	Endovascular repair 16 × 5.5-mm Zenith endograft, complicated by common femoral artery thrombosis, 24 d follow-up
Khanna [25]	2007	7/M	Focal infrarenal aortic intimal flap	Aspirin only, intimal flap healed at 2 mo of follow-up
McCarthy [26]	2007	10/F	Infrarenal aortic thrombosis, paraplegia, compartment syndrome	10-mm interposition PTFE graft, residual paraplegia at 8 y of follow-up
Anderson [27]	2008	7/F	NR	Primary repair
		7/M	NR	Non-operative
Burjonrappa [28]	2008	9/M	Focal dissection of the infrarenal abdominal aorta	Non-operative Stable at 18 mo of follow-up
Heck [29]	2009	1.3/M	Pseudoaneurysm	Non-operative, stable at 3 mo of follow-up
Blanco [30]	2011	2/M	Aortic bifurcation rupture	Primary repair with Prolene (Ethicon) sutures, stable at 6 mo of follow-up
Sadaghanloo [31]	2011	4/B	Pseudoaneurysm	Primary repair non-absorbable interrupted suture on d 8, no follow-up
		7/F	Pseudoaneurysm	Primary repair non-absorbable interrupted suture on d 10, no follow-up
Shalhub [32]	2012	6/NR	Infrarenal abdominal aortic rupture	Primary repair with Prolene sutures, postoperative death
Papazoglou [33]	2015	9/M	Infrarenal abdominal aortic dissection, paraplegia	Abdominal aorta 12 × 40-mm self-expandable Protégé Everflex (ev3) stent Iliac 8 × 60 mm, Protégé Everflex, 1-y follow-up, persistent paraplegia
Parrish [34]	2015	12/M	Abdominal aortic intimal flaps	Non-operative, pseudoaneurysm at 1 y of follow-up
Daniele [35]	2017	7/M	Pseudoaneurysm	Bovine pericardium patch, no follow-up
Jammeh [36]	2020	2/NR	Renal artery avulsion, aortic intimal flap	Right nephrectomy, stable at 2 y follow-up
		11/NR	Contained transection of the juxtarenal aorta, paraplegia	12-mm Dacron (Dupont) interposition, paraplegia at 15 mo

Abbreviations: F, female; M, male; NR, not reported; PTFE, polytetrafluoroethylene.

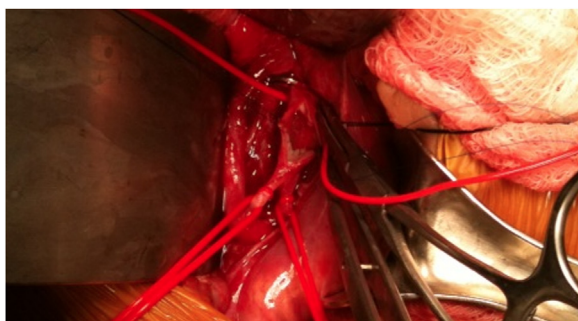


Fig. 1 – Transabdominal exposure of a left common iliac artery in a 5-year-old child post blunt injury to the abdomen, revealing the intimal transection and repair primarily.

energy trauma [8]. Diagnosis of vascular injury depends on identifying hard signs of vascular injury (eg, external bleeding and expanding hematoma) and a high level of suspicion based on the trauma mechanism. Arterial injury diagnostic strategies are variable among the different centers, including angiography and duplex examination [37]. Duplex ultrasound can be used to evaluate extremity arterial injuries in children, and axial imaging is necessary to evaluate neck, chest, and abdominal injuries. Arterial vasospasm is frequently encountered in the pediatric population and is associated with fractures or a cold extremity. Severe vasospasm can present with a pulseless extremity. Thus, warming the patient and critically evaluating the etiology of the pulseless limb is imperative [13]. Exsanguinating extremity bleeding can be controlled with prehospital tourniquet application, a practice that has shown benefits [38]. When extremity injury is associated with a fracture, the first line of management is the closed reduction and fixation of the fracture. Arterial management depends on the degree of associated ischemia and immediate surgical repair of the artery is indicated in clear ALI cases. An area of variation in practice exists in the cases of brachial artery injury associated with supracondylar humeral fracture. In a systematic review of 16 studies and 608 cases of pediatric brachial artery injury associated with supracondylar humeral fracture, 57.4% regained pulses after the reduction and fixation of the humerus, and 42% remained pulseless (referred to as “pink pulseless hand”) [37]. Of the patients with a pink pulseless hand, variation in practice exists, with nearly one-half (48.6%) undergoing surgical management with brachial artery exploration and the remaining 51.4% were managed non-operatively with a “watchful expectancy” approach [37]. The latter is dictated by the brachial artery size and feasibility of repair.

The predominant vascular repair modality in pediatric surgery remains open surgical repair [7,9,39]. Similar to adults, surgical repair principles include arterial exposure and restoration of blood flow to end organs. This can be achieved via primary repair, end-to-end anastomosis, interposition grafting, and bypass grafting. As with adults, autogenous vein graft is preferred over prosthetics in extremity arterial bypasses [13]. Challenges unique to the pediatric population include the small diameter of the arteries and the

need to accommodate future axial and radial arterial growth. Interrupted sutures during anastomoses creation allow the repaired artery to grow as the child grows without stenosis [13,40]. Another unique challenge in this population is the high degree of arterial spasticity leading to vasospasm with trauma. During arterial repairs or cannulation procedures, intraoperative papaverine can be used as an adjunct to promote the relaxation of arterial smooth muscle and resolve vasospasm [41].

The role of endovascular techniques in the pediatric population is evolving. The main challenge to adoption is devices too large for pediatric vessels. In addition, pediatric arteries are prone to iatrogenic dissection and vasospasm. With the development of smaller-caliber devices, there has been an increase in the prevalence of endovascular techniques in surgical trauma care for children in recent years. A review of the 2007–2014 NTDB of 35,771 children with vascular injuries demonstrated an increase in endovascular techniques in managing blunt trauma from 7.8% to 12.9% [39], most commonly, thoracic endograft deployment and internal iliac embolization [39]. Children with high Injury Severity Scores (ISS) had a higher proportion of endovascular procedures than those with lower ISS [39]. Children who underwent endovascular procedures were older than those who underwent open operations (mean \pm SD age, 12.4 ± 4 v 10.3 ± 4.8 years, respectively) [39]. This is likely related to the size of arterial access and target vessels’ inability to accommodate existing sizes of endovascular devices such as thoracic endografts. A major consideration in using thoracic endografts is future axial and radial aortic growth with the possibility of developing coarctation physiology due to the stable device size compared with the growing aorta. These biological demands should shape future biomaterials and endovascular stent graft designs. Literature on the use of thoracic aortic endografts in pediatric trauma patients is limited to case reports [42]. The youngest patient with a thoracic aortic injury treated was 8 years old [42].

2.2. Pediatric access-related acute limb ischemia

In infants and children, iatrogenic complications from femoral cannulation for invasive hemodynamic monitoring, extracorporeal membrane oxygenation, or cardiac catheterization are the leading cause of ALI [43–47]. The highest risk for catheterization and intervention-related ALI is observed in premature infants and children with congenital heart disease [43,45–47]. Arterial thrombosis occurs due to intimal injury related to access. Other complicating factors include increased blood viscosity in this population, stasis due to low cardiac output or vasospasm, and small arterial diameter compared with catheter size [48]. Vasospasm increases wall contact surface area with the cannula and is associated with fibrin deposition and thrombosis [48]. The most common injury sites include the external iliac artery (42%) and the common femoral arteries (30%) [49].

Management includes systemic anticoagulation and duplex ultrasound surveillance [45,47,48]. In a case series by Warner et al [45], of 32 patients (59% were male) with lower extremity arterial thrombosis, limb preservation was 100% and no surgical interventions were required. Follow-up duplex ex-

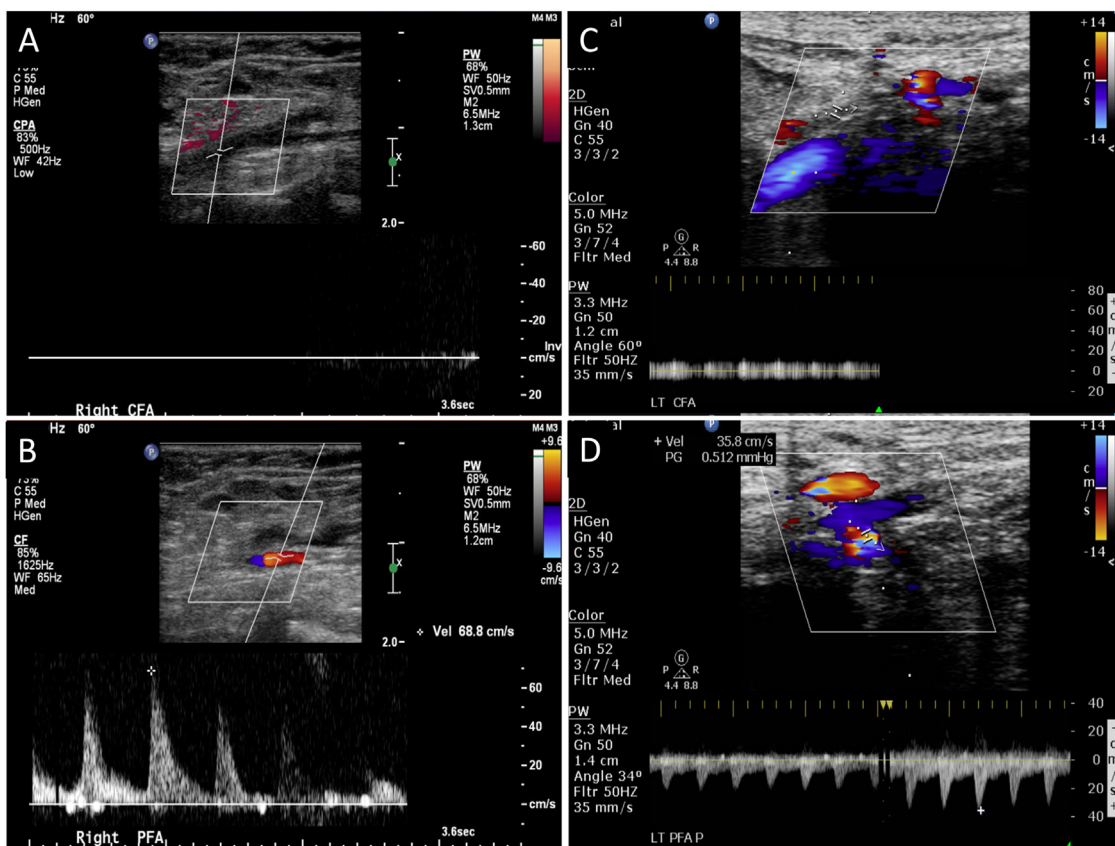


Fig. 2 – (A) Arterial duplex ultrasonography of an 8-month-old male infant demonstrating the absent flow in the right common femoral artery. (B) The flow in the profunda femoris is antegrade beyond the thrombosed common femoral artery. (C) Arterial duplex ultrasonography of a 7-month-old male infant with a thrombosed left common femoral artery. (D) The flow in the profunda femoris is retrograde beyond the thrombosed common femoral artery.

amination demonstrated arterial recanalization in 57% of the cases. In patients with occluded common femoral arteries, the flow was antegrade in the profunda femoris artery in 64%, and retrograde in 31% of duplex ultrasound studies (Fig. 2). In another study by Kayssi et al [49], of 151 patients (55% were male), the non-operative strategy was successful in 93.4% of the cases. Rapid arterial collateralization is likely the reason for successful limb preservation without vessel recanalization [47,49]. Thrombolysis is used in cases where anticoagulation is ineffective, and there are no contraindications for its use [48].

Surgical intervention for access-related ALI includes surgical thrombectomy, arterial repair, fasciotomy, and amputation [43,48,49]. According to a recent study that examined the Japanese national administrative claims data and discharge records for 948 pediatric patients (55% were male) who received venoarterial extracorporeal membrane oxygenation treatment, the incidence of fasciotomy and amputation was found to be 0.6% and 0.7%, respectively [43]. In Kayssi et al's [49] study, a 6.6% incidence of operative interventions was reported. The interventions included thrombectomy, bypass, arterial reconstruction, and fasciotomy. In general, operative interventions in neonates and children younger than 3 years have worse outcomes than older children, and consequently, non-operative management is preferred in this population [48].

2.3. Pediatric aortic dissection and aneurysms

AD and aneurysms rarely occur in children. An examination of the 1996–2005 Statewide Planning and Research Cooperative System database of New York State revealed that out of 12,142 cases of AD, only 45 (82% were in male patients) occurred in individuals 21 years or younger, and none were younger than 15 years [50]. In this cohort, the etiology was most commonly trauma (19 of 45 cases), followed by Marfan syndrome (MFS; 11 of 45 cases) [50]. Analysis of the Kids' Inpatient Database, a national sample of pediatric admissions in the United States between 1997 and 2009 identified 168 cases of AD (76% were in male patients) with the following age distribution: 18 (11%) were younger than 1 year, 38 (22.6%) were aged 1 through 14 years, and 112 (67%) were aged 15 through 19 years [51]. The study interestingly demonstrated regional variation, with 42% of the AD cases occurring in the South [51]. The Pediatric Health Information System database, a multi-institutional administrative database of more than 40 participating pediatric hospitals, evaluated all index cases of aortic patients younger than 30 years between 2004 and 2011. The incidence of AD was 3 in 100,000 pediatric hospitalizations, with a bimodal age distribution of 0 through 5 years (27%) and 15 through 20 years (43%) [52]. The most commonly associated condition with AD was congenital heart disease (38%), followed by trauma (24%)

Table 2 – Medical history and management of aortic dissection in pediatric aortic dissection case reports.

First author	Year	Age/sex	Type of dissection	Medical history
Hibino [53]	2006	3 y/M	A	Atrial septal defect, non-operative management
Eun [54]	2011	11 y/M	B	IgA nephropathy, non-operative management with plan for repair
Morais [55]	2011	15 y/M	A	VEDS
Ware [56]	2014	16 y/M	A	ACTA2, valve-sparing aortic root replacement, ascending and arch replacement
		17 y/M	A	ACTA2, valve-sparing aortic root replacement, ascending and arch replacement
Besli [57]	2015	14 y/M	B	Familial aortopathy, transfer initiated to outside hospital for surgical intervention
Regalado [58]	2014	14 y/M	B	ACTA2
		14 y/M	B	ACTA2
		14 y/M	B	ACTA2
		17 y/M	A	ACTA2
Shalhub [59]	2019	16 y/M	B	PRKG1, TAAA2 repair 2 y after TBAD
		16 y/M	B	PRKG1, TAAA2 repair 1 y after TBAD (5.5-cm DTA)
D'Addese [60]	2019	17 y/F	A	Prior heart transplantation 2 ×, known ascending aortic dilation, homozygous mutation of MYBPC3 gene, emergency ascending aorta and arch replacmenet with Dacron (Dupont) graft
Venardos [61]	2020	3 d/F	B	Hypoplastic left heart syndrome, aortic and mitral atresia, repaired with pulmonary allograft patch
Mamishi [62]	2021	14 y/M	A	Williams syndrome, deceased on arrival to hospital
Comentale [63]	2021	11 mo/M	A	LDS, David V procedure with 18-mm Dacron graft
Dueppers [64]	2022	9 y/M	B	LDS, nederal management, staged repair for rapid degeneration with David procedure with frozen elephant trunk, then thoracoabdominal repair with 20-mm Dacron interposition graft to supramesenteric aorta with celiac reimplantation
Liang [65]	2021	10 y/F	A	Bicuspid aortic valve, replacement of aortic valve with St. Jude mechanical valve, aortic root, ascending aorta, and arch
Matsushita [66]	2021	11 y/M	B	FBN1 and TGFBR2, medical management required open descending aortic repair with partial cardiopulmonary bypass on hospital day 5 for enlargement of the false lumen
Rathnayake [67]	2022	15 y/M	A	Autism Bentall procedure

Abbreviations: DTA, XXXX; LDS, Loeys-Dietz syndrome; MFS, Marfan syndrome; TAAA, thoracoabdominal aortic aneurysm; TBAD, type B aortic dissection; VEDS, vascular Ehlers-Danlos syndrome.

and connective tissue disease (16%) [52]. Similar to the New York State and the Kids' Inpatient Database data, most affected individuals were male (69%) [52]. Table 2 summarizes the case reports of AD in the pediatric population [53–67].

Risk factors for AD in children include male sex; hypertension; genetic aortopathy; congenital abnormalities, such as aortic coarctation; and trauma [50–52]. Pathogenic variants in genes involved in the transforming growth factor (TGF)– β pathway and smooth muscle proteins can cause genetic aortopathies or heritable aortopathies [68].

Syndromic genetic aortopathies include MFS, Loeys-Dietz syndrome (LDS), and vascular Ehlers-Danlos syndrome (VEDS). MFS is due to autosomal dominant inheritance pathogenic variants in *FBN1* affecting 1 in 5,000 individuals. LDS is due to autosomal dominant inheritance pathogenic variants in *TGF- β receptor 1* (*TGFBR1*, *TGFBR2*, *SMAD3*, *TGFB2*, and *TGFB3*) [68–70]. MFS accounted for 24% of AD cases younger than 21 years in the Statewide Planning and

Research Cooperative System database, 14% in the Kids' Inpatient Database study, and 12% in the Pediatric Health Information System study [50–52]. In the Genetically Triggered Thoracic Aortic Aneurysm and Cardiovascular Conditions (GenTAC), a well-characterized registry of pediatric and adult individuals with genetic aortopathies, there were 11 of 245 (4%) operations for AD in the pediatric population: 5 with MFS, 3 with LDS, and 3 with non-syndromic heritable thoracic aortic aneurysm [71]. Low numbers in pediatrics are attributed to early surgical intervention in a closely followed population [71]. A known risk factor for ascending thoracic AD (type A) in individuals with MFS and LDS is a dilated aortic root; however, this population is also at risk for descending thoracic AD (type B). Vertebral artery tortuosity is a characteristic feature of patients with MFS and LDS. Morris et al [72] quantified the degree of tortuosity as the ratio of the length of the vertebral artery along its course (actual length) to the longitudinal distance the vessel travels in space (straight length), as demonstrated

in Fig. 3. A higher vertebral artery tortuosity (calculated as $[\text{actual length} / \text{straight length} - 1] \times 100$) was associated with a younger age at first AD in patients with MFS and LDS [72].

VEDS is due to autosomal dominant inheritance pathogenic COL3A1 affecting 1:50,000 individuals. VEDS accounts for 5% of EDS (which includes 14 subtypes) [73]. Children with VEDS are frequently born more prematurely than the general population, which is attributed to premature rupture of members due to defective type III collagen production [74]. Adolescents are at increased risk for AD, aneurysms, and sudden aortic and arterial rupture (Fig. 4), intestinal perforation (most commonly the sigmoid colon), and spontaneous pneumothorax [75–77]. A recent survey of individuals living with VEDS demonstrated significant frustration with the lack of VEDS-specific knowledge among emergency physicians [78]. This can translate to a delay in aortic or arterial dissection or rupture when presenting to the emergency department.

Pathogenic variants in smooth muscle cell proteins, such as ACTA2, MYH11, MYLK, and PRKG1 are associated with non-syndromic genetic aortopathy. This population is also at risk for AD (Fig. 5) [58,59,79].

It is important to note that there are significant differences in the cumulative risk of aortic aneurysm and AD among individuals with different genotypes [79]. This improved understanding of genotype–phenotype correlation guides elective root and ascending aortic repair recommendations [80]. We anticipate that in the future, personalized guidelines can be developed for the remainder of the aorta and arteries [80].

Unlike the syndromes caused by genetic aortopathy, Turner syndrome is caused by monosomy for the X chromosome during embryonic development and has an estimated prevalence of 1:2,000 live births. Turner syndrome is also a risk factor for AD. Although the mean age of AD was 30.7 years among 85 cases reported between 1961 and 2006, the youngest was 4 years old [81].

Congenital abdominal aortic aneurysms without a diagnosable etiology are also rare and have been described in the pediatric population in the form of case reports. Repair is technically challenging in infancy and childhood due to the small aortic size and potential for the child's growth [82]. There are no standard approaches to repair, and conduit choices include cryopreserved allografts and prosthetic grafts (Dacron [Dupont] and Gore-Tex [W. L. Gore and Associates]). Most case reports have short-term follow-up. Salient technical points to consider in these cases is that patency of grafts at diameters < 6 mm is generally poor and that the child will likely require a redo operation as they grow [82].

3. Emergency vascular surgical care during pregnancy

“Complicated pregnancy” ranked as the sixth leading cause of death among female patients aged 15 through 35 years in the United States from 2000 to 2020 [4]. Several cardiovascular changes occur during pregnancy, and these can be associated with some of the vascular emergencies that occur during pregnancy. These include increased circulating blood volume and cardiac output, which peak in the third trimester. More-

over, circulating pregnancy hormones, namely relaxin, estrogen, and progesterone, adversely affect the structural integrity of the arterial wall. The increase in arterial elasticity induced by these hormones may be amplified by fragmentation of the internal elastic lamina, subendothelial thickening, medial fibrodysplasia, and glycosaminoglycan deposition in the subintimal and medial layers [83]. Both estrogen and progesterone increase significantly in the third trimester at 30 to 32 weeks of gestation and progress with gestational age. The progression peaks in the late third trimester around the time of delivery. In addition, pregnancy is a prothrombotic state, thus increasing the risk for pulmonary embolism and deep vein thrombosis (DVT).

Overall, vascular emergencies requiring vascular surgical consultations or interventions are rare in pregnant individuals. These include vascular injuries, ALI, AD, visceral aneurysm rupture, and limb-threatening thromboembolic complications. When these emergencies occur, multidisciplinary team care is essential for patient care and should include fetal assessment and monitoring.

3.1. Vascular emergency imaging concerns during pregnancy

For most vascular emergencies during pregnancy, computed tomography is the diagnostic modality of choice to expedite accurate diagnosis and surgical intervention planning. Hesitancy to perform a computed tomography scan because of concerns about fetus radiation exposure can lead to delays in diagnosis [84]. Although it is important to decrease the exposure to ionizing radiation when possible, the ionizing radiation dose in most imaging modalities is much lower than the exposure associated with harm, especially in the second and third trimesters (usually below the threshold of < 50 mGy) [85]. The mean radiation dose to patients undergoing thoracic endovascular aortic repair is 323.7 ± 161.0 Gy/cm², equivalent to 45.3 ± 22.5 mSv of uterine radiation dose (organ-specific) [86,87].

3.2. Traumatic vascular injuries during pregnancy

Traumatic injury is the leading cause of non-obstetric maternal death, affecting 7% to 10% of pregnancies [88]. The leading causes of trauma during pregnancy are MVC, falls, and domestic violence [89]. Pregnancy complications related to blunt trauma include uterine rupture, placental abruption, preterm birth, perinatal death, and cardiac arrest [89]. Although specific outcomes related to major vascular injuries have not been well described, trauma care principles are similar to non-pregnant individuals, with the goal of maternal stabilization.

3.3. Acute limb ischemia in pregnancy

ALI during pregnancy and the postpartum period is rare. A systemic review of 14 articles found 14 patients (median age, 31.5 years). Eight of the patients analyzed had an embolic origin, with peripartum cardiomyopathy being the primary cause. Other causes included popliteal entrapment, iatrogenic factors, and polyarteritis. Management was surgical embolectomy in 11 cases and anticoagulation alone in 3 cases. None

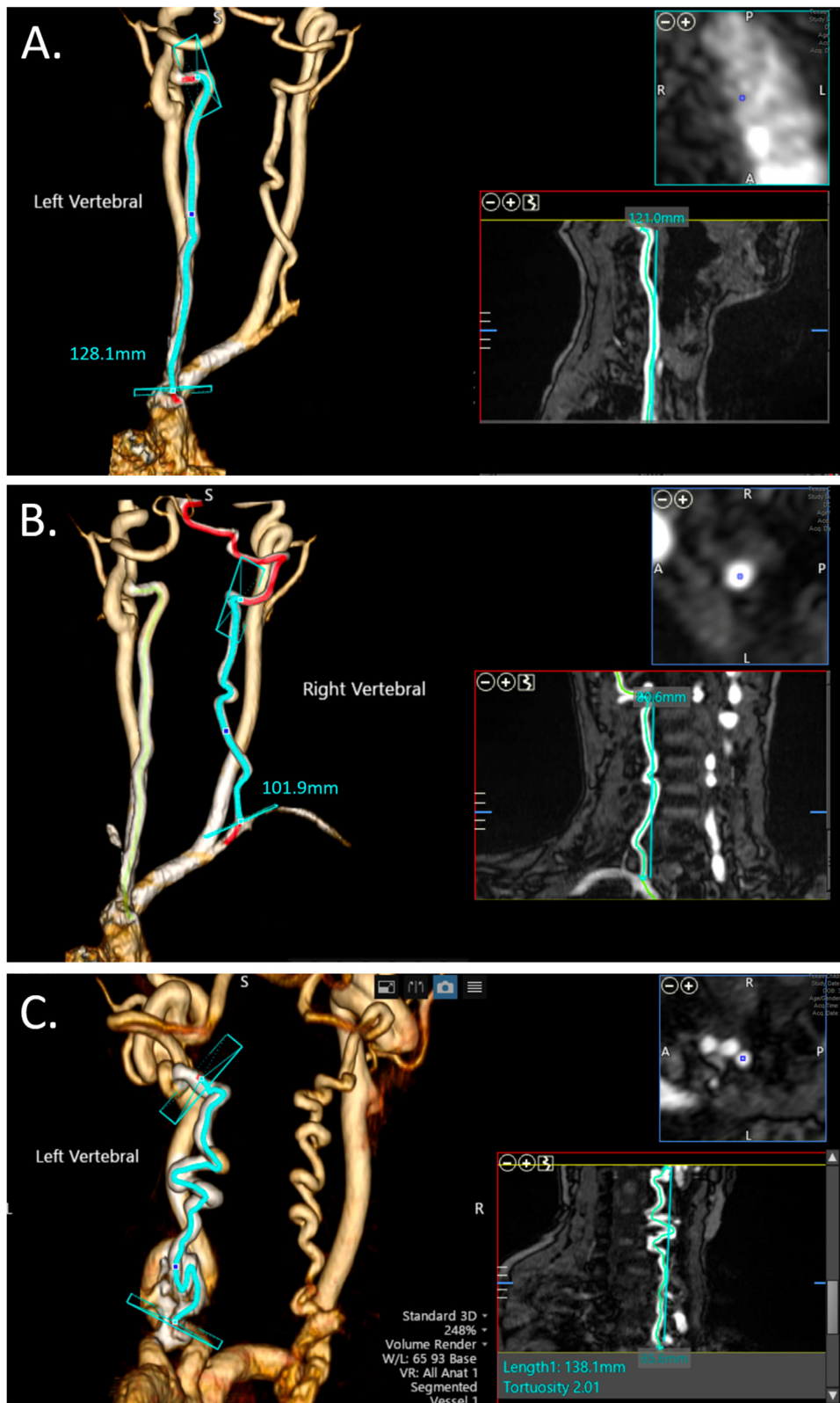


Fig. 3 – Measuring vertebral artery tortuosity index (VTI). The curved length and straight (crow's flight) length are measured in three-dimensional space. VTI for each vessel is calculated using the equation: $VTI = ([\text{curved length} / \text{straight length}] - 1) \times 100$, and the maximum of the two VTIs is the study VTI. (A) left vertebral artery VTI in patient 1 = $([128.1 / 121.0] - 1) \times 100 = 5.9$. (B) right vertebral artery VTI in patient 1 = $([101.9 / 80.6] - 1) \times 100 = 26.4$. Patient 1: VTI = 27.4. (C) Left vertebral artery VTI in patient 2: $VTI = ([138.1 / 45.9] - 1) \times 100 = 201$. Figures courtesy of Shaine A. Morris, MD, Division of Cardiology, Department of Pediatrics, Baylor College of Medicine, Texas Children's Hospital, Houston, TX.

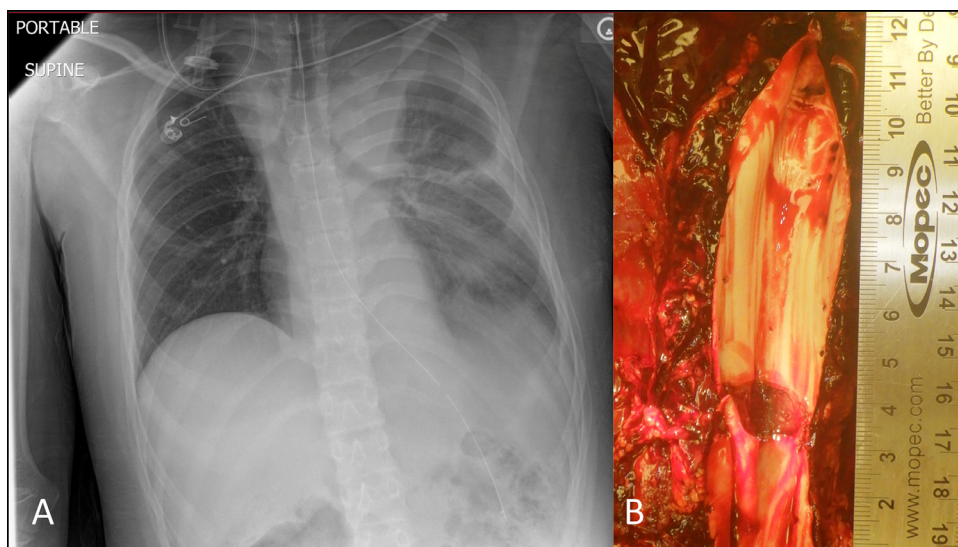


Fig. 4 – (A) Chest x-ray of a 15-year-old boy with vascular Ehlers-Danlos syndrome presenting with sudden atraumatic onset back pain demonstrating a widened mediastinum. The patient became hypotensive and had a cardiac arrest. (B) The autopsy demonstrated full-thickness rupture of the descending thoracic aorta at T9 to T10 with dissection to the level of the aortic bifurcation.

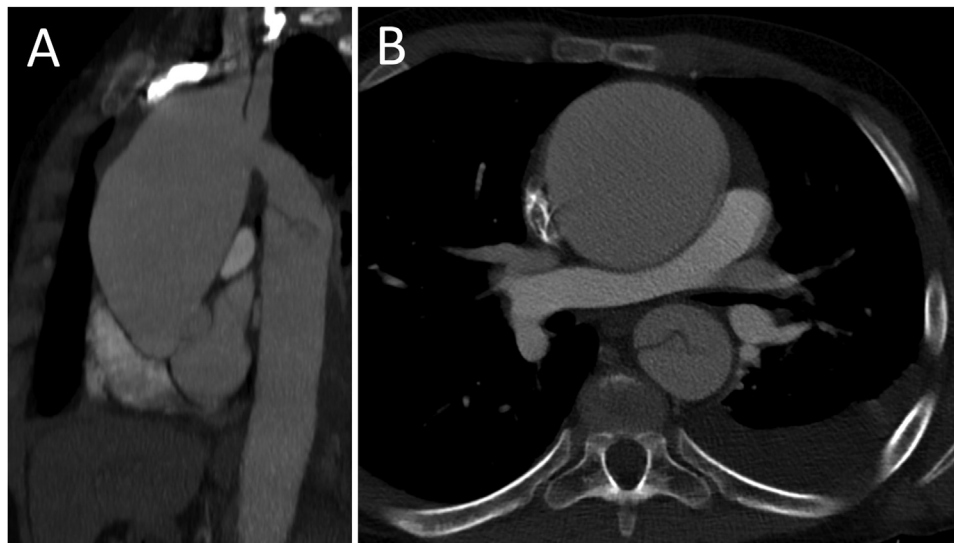


Fig. 5 – (A) Sagittal and (B) axial images of a computed tomography scan of a 14-year-old boy with a large ascending aortic aneurysm and a type B aortic dissection related to a pathogenic variant in ACTA2.

of the patients underwent an endovascular intervention [90]. Heparin is the anticoagulant of choice in this population, as it does not cross the placenta. Low-molecular-weight-heparin is preferred over unfractionated heparin. Warfarin is contraindicated during pregnancy due to the risk of teratogenicity; however, it can be administered postpartum. Direct oral anticoagulants are avoided during pregnancy and postpartum due to a lack of safety information [91].

3.4. Aortic dissection in pregnancy

AD during pregnancy is a rare but catastrophic event estimated to occur in 0.1% of all AD cases in the United States

and 0.0004% of all pregnancies [92]. AD is the third most frequent cause of maternal death from cardiovascular disease and is associated with 12% to 16% maternal mortality and 28% to 40% fetal mortality [92–95]. Most pregnancy-related ADs occur in the third trimester [93,96,97]. Another vulnerable time is the postpartum period [93,96,97]. Analysis of the 1998–2019 International Registry of Acute Aortic Dissection data demonstrated that 29 individuals had an acute AD during pregnancy or postpartum (< 12 weeks, thus accounting for 0.3% of 9,709 individuals with AD and 1% of 2,788 women with AD who had pregnancy data recorded. AD was type A in 45% and type B in 55% of the cohort. Timing of AD was most common in the third trimester and in the postpartum

period at a mean interval of 12.5 days (range, 2 to 56 days) postpartum [97]. In a single-institutional series from the University of Texas at Houston, among 1,190 ADs, 10 individuals had peripartum AD (0.8%) at a median age of 31.5 years (interquartile range, 28 to 38 years): 5 type A and 5 type B ADs [98]. In addition to the cardiovascular physiologic changes that occur in pregnancy, gestational hypertension, such as seen with preeclampsia and eclampsia, may increase the risk of AD [94].

Pregnant individuals with genetic aortopathy are at high risk for AD [97]. MFS is the most common genetic aortopathy present in AD in pregnant individuals. Additional genetic aortopathy risk factors include VEDS, LDS, and non-syndromic heritable thoracic aortic disease [99]. AD is estimated to occur in 5% to 7% of individuals with MFS and LDS [100]. Among the International Registry of Acute Aortic Dissection cohort, 18 had genetic aortopathy [97]. Among the University of Texas Health Science Center at Houston cohort, 3 of 10 had MFS [98]. In the National Heart, Lung, and Blood Institute GenTAC cohort, data were available for 94 individuals with 227 pregnancies (mean maternal age, 29 years; range, 13 to 43 years). Among this cohort, 7 (7.4%) experienced an AD: 4 type A and 3 type B ADs. Most of the ADs (n=5) occurred in the postpartum period [100]. Sub-analysis revealed that pregnancy in individuals with MFS was associated with an eightfold increase in the risk of AD compared with non-pregnant individuals. Pathogenic variants in smooth muscle cell proteins associated with non-syndromic genetic aortopathy, such as *ACTA2*, *MYH11*, *MYLK*, and *PRKG1*, are risk factors for AD. Among individuals with *ACTA2* pathogenic variants, 20% had AD associated with pregnancy [58]. Turner syndrome and bicuspid aortic valve are also risk factors for AD during pregnancy [84].

The most recent American Heart Association guidelines recommend shared decision making for individuals with known genetic aortopathy when considering the cardiovascular risks of pregnancy, the diameter thresholds for prophylactic aortic root surgery to prevent the risk of type A AD, and the mode of delivery [101]. Generally, among individuals with MFS, LDS, or bicuspid aortic valve with dilated aortic roots or ascending aorta, prophylactic aortic aneurysm repair is recommended before conception, with the threshold personalized by the specific gene [101].

Treatment of AD during pregnancy varies by type [84]. In the recently reported International Registry of Acute Aortic Dissection dataset, all 13 patients with type A AD underwent surgical repair, most with type B (n=10) were treated medically, and the remainder were treated with endovascular procedures (n=3) and surgical repair (n=3) [97]. In our experience, we prefer medical management for type B AD as long as it is not associated with malperfusion or rupture with planned repair in the postpartum period (Fig. 6). Type A AD requires emergency repair regardless of gestational age. The timing of surgical aortic repair versus delivery depends on gestational age and AD complications. When it occurs at 28 weeks' gestation, the emergency aortic repair is performed with fetal monitoring and modification to anesthesia and cardiopulmonary bypass in an attempt to decrease the fetal risk by maintaining maternal blood pressure, cardiac output, hemoglobin, and pH [87,93,102,103]. When type A AD occurs at a gestation age of >



Fig. 6 – Sagittal images of a computed tomography scan of a female patient with Marfan syndrome who presented with a type B aortic dissection in the third trimester of pregnancy.

28 weeks, a cesarean section delivery is performed, followed by aortic repair [87,93,97,101,103].

3.5. Splenic artery aneurysms in pregnancy

Visceral arterial aneurysms are uncommon in the general population and even rare in pregnant patients, with a reported incidence of 0.16% to 0.78% [104]. In both populations, SAAs are the most common. Pregnancy hormonal and physiological changes likely play a role in SAA development. As many as 50% of ruptures may be associated with pregnancy; in this setting, the maternal and fetal mortality rates approach 75% and 95%, respectively [83,104-106]. Diagnosis in these cases can be challenging and can be delayed, as presenting signs and symptoms may mimic many obstetric problems. Although the intervention of SAA is recommended for any non-ruptured aneurysm > 3 cm, current guidelines support treatment of SAA of any size in women of childbearing age, regardless of pregnancy status due to the risk of rupture [105]. Both open and minimally invasive techniques (mostly transcatheter em-

bolization) may be used, depending on the anatomic features of the SAA [105].

3.6. Emergent thromboembolic complications in pregnancy

Pregnant individuals have a fivefold risk of DVT and 15% of maternal mortality is caused by pulmonary embolism [107–109]. There have been no randomized trials of thrombolytic agents in pregnant patients for any indication, and only a handful of studies have documented their use. A literature review summarized 141 cases of systemic thrombolysis in pregnant patients for stroke, DVT, pulmonary embolism, and mechanical valve prosthesis thrombosis, in which there were only four maternal deaths and two fetal losses, none related to complications of thrombolysis [110]. These studies are heterogeneous, but indicate a role for thrombolysis during pregnancy in life-threatening situations. Generally, radiation exposure is to be minimized by shielding the abdomen with a lead apron when possible, reducing the pulse fluoroscopy frame rate during the intervention, and minimizing digital subtraction angiography [111]. Phlegmasia cerulea dolens is a rare complication of proximal DVT. In addition to systemic anticoagulation, catheter-directed thrombolysis (CDT), pharmacomechanical thrombolysis, or operative venous thrombectomy may be indicated, depending on the characteristics of the thrombus and the patient's response to anticoagulation [111]. Stenting of the common and external iliac veins with self-expanding stents may be needed and can be assessed on a case by case basis [111]. Three case reports described the successful use of CDT in patients in the first trimester of pregnancy without pregnancy-related complications [107–109]. Ladha et al [108] described a case of iliofemoral DVT complicated by phlegmasia cerulea dolens during a first-trimester pregnancy treated with a operative venous thrombectomy in addition to CDT and pharmacomechanical thrombolysis, followed by common iliac stent placement. To avoid ionizing radiation, Dua et al [107] described the use of intravascular ultrasound instead of angiography for CDT with recombinant tissue plasminogen activator for a similar case.

4. Emergency vascular surgical care in the frail population

A significant portion of patients undergoing emergent vascular operations are older adults. Increasing age is associated with an increased risk of morbidity and mortality during urgent and emergent vascular operations [112]. However, the ability to withstand the physiologic stress of a vascular emergency is variable and is not entirely age-dependent. Relying solely on age may lead to overestimating or underestimating a patient's overall physiologic reserve. For instance, a physically active individual in their 90s may be more physiologically resilient than a person 2 decades younger, but is frail. Thus, surgical decision making must also consider an individual's frailty and wishes for end-of-life care options. Numerous studies have demonstrated that frailty increases postoperative morbidity and mortality risk [113–115]. A recent meta-analysis demonstrated a fourfold increase in 30-day mortality

and a twofold increase in long-term mortality among frail individuals after vascular surgery [116].

Frailty is a biological syndrome of “decreased reserve and resistance to stressors, resulting from cumulative declines across multiple physiologic systems, and causing vulnerability to adverse outcomes” [117]. The term was first described by Vapuel in 1979. Subsequently, Fried and colleagues [117] introduced the “frailty phenotype” concept in 2001. The phenotype evaluates five domains, of which patients with three or more are considered frail. These domains are unintentional weight loss, slow gait speed, self-reported exhaustion, impaired grip strength, and low physical activity [117]. An estimated 25% to 50% of older surgical patients are living with frailty [118]. The tools are available to evaluate patients' physiologic reserve and stratify their surgical risk. The time needed to complete each tool varies widely. In circumstances of emergency vascular surgery, quick assessment of frailty is imperative, as it would facilitate shared decision making with the patient, if able, and their caregivers.

Gottesman and McIsaac [118] suggest using the Canadian Study of Health and Ageing Clinical Frailty Score, the Risk Assessment Index, and Frailty Index tools in the acute care surgery setting due to their ease of use [118]. The Clinical Frailty Score is an 8-point ordinal scale that takes less than 1 minute to complete [119]. The Clinical Frailty Score uses visual cues and brief vignettes to summarize a patient's health and activity level, stratifying them on a frailty continuum [119]. The Risk Assessment Index uses 14 domains of multifactorial deficits to similarly stratify a patient's frailty on an 81-point scoring system. This can be completed in 2 minutes if information technology is used. The frailty index assesses 30 domains of the patient's status, adding the presence of deficits into a cumulative score. This tool takes 5 to 10 minutes to complete, which may render it too cumbersome to be used in an emergency setting. Although multiple tools have been used to evaluate frailty in vascular surgery, the use is variable by practice [116].

4.1. Palliative care

The delineation of end-of-life care wishes includes using advanced directives and physician orders for life-sustaining treatment forms [120]. Patients with vascular emergencies often have complex medical histories, which, combined with the time-sensitive nature of the emergency setting, can make it challenging to discuss their care goals.

There has been increasing recognition of the value of palliative care expertise in vascular surgery. The term *palliative care* was coined by Balfour Mount, a surgeon, in the 1970s [121]. Implementing palliative care in surgical patients has been recognized as a service that improves patient quality of life and patient-physician communication. In addition, it reduces health care costs [122]. The American College of Surgeons and the Robert Wood Johnson Foundation formed a workgroup in 2003 to address palliative care applications in surgical settings [121]. The workgroup identified the following seven domains as potential research areas: (1) surgical decision making, (2) patient decision making, (3) end-of-life decision making, (4) symptom management, (5) communication, (6) processes of care, and (7) surgical education about palliative care [121].

There are limited data on palliative care utilization in patients undergoing emergency vascular operations. The Healthcare Cost and Utilization Project's Nationwide Inpatient Sample data from 2009 to 2014 was analyzed to evaluate the role of palliative care in the care of patients with ruptured abdominal aortic aneurysms [123]. Diagnostic codes for palliative care were used to determine utilization in 28,255 patients identified in the study. Among the cohort, only 14% received a palliative care consultation, which was more frequent among patients who did not undergo operative repair (41% v 6.3%) [123]. Female patients and patients 80 years or older were more likely to receive a consultation, with regional variation showing higher utilization rates in the Western and Southern regions of the United States [123]. A noteworthy finding was that patients who received palliative care consultation had significantly lower overall health care costs and shorter hospital stays [123].

Palliative care utilization was also evaluated among individuals with chronic limb-threatening ischemia. A 2021 study by Kwong et al [124] of 111 patients (68.5% were male, 26.1% were Black) who underwent a below-the-knee amputation found that only 3 received a palliative care consultation, and 6 received palliative care consultation before death. A 2022 study by Morton et al [125] of 292 patients with chronic limb-threatening ischemia (61% were male, 53% were Black) who underwent a major amputation found that 12% received a palliative care consultation during the hospitalization when they underwent the amputation. The median time to the consultation was 6 days (interquartile range, 1 to 17 days) after the amputation. The patients who did receive a palliative care consult were more likely to be discharged to hospice than to die in the hospital [125].

5. Conclusions

The pediatric, pregnant, and frail populations have unique vascular emergency needs and physiological characteristics. Understanding the epidemiology of vascular emergencies in these populations is essential to diagnose and manage these conditions effectively. Considering each population's unique characteristics is crucial to the emergent vascular surgical interventions decision making. Collaborative and multidisciplinary care is vital in managing these special populations and achieving optimal patient outcomes.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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