Aortopathy in Adults with Congenital Heart Disease

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Abstract

Over the past two decades, there has been an increasing body of data that has acknowledged the progressive nature of aortic dilation in adult patients with congenital heart disease. Aortic disease can have overwhelming effects on affected individuals, resulting in severe morbidities including aneurysms, tears, dissections, and aortic valve regurgitation secondary to annular dilation. Several explanations have been provided for this rising paradigm of aortic disease including the incremental number of adult survivors with moderate and complex forms of congenital heart defects, along with an associated improvement in diagnostic cardiovascular imaging modalities like cardiac magnetic resonance and computed tomographic angiography. Although the natural history of aortic root dilation in patients with connective tissue disorders like Marfan syndrome has previously been described, the natural history of the aorta in adult congenital heart disease remains poorly assessed. This review describes the presence of "aortopathies" in adults with the more common congenital heart defects including bicuspid aortic valve, repaired Tetralogy of Fallot, conotruncal lesions, and patients with single ventricular physiology after Fontan palliation.

Keywords

Adults with congenital heart disease • Aortic dilation • Aortic regurgitation • Aortopathies • Bicuspid aortic valve • Congenital heart disease • Congenitally corrected transposition of the great arteries • Conotruncal defects • d-Transposition of the great arteries • l-Transposition of the great arteries • Tetralogy of Fallot • Truncus arteriosus

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Abbreviations	
ACHD	Adults with congenital heart disease
AD	Aortic dilation
AR	Aortic regurgitation
BAV	Bicuspid aortic valve
cc-TGA	Congenitally corrected transposition of the great arteries
CHD	Congenital heart disease
d-TGA	d-transposition of the great arteries
l-TGA	Transposition of the great arteries
TA	Truncus arteriosus
TOF	Tetralogy of Fallot

Introduction

The aorta is the main trunk of a series of vessels which convey oxygenated blood to the tissues of the body. It commences at the superior part of the left ventricle, where in adults it measures about 3 cm in diameter. It then continues to ascend for a short distance, arches backward and to the left, over the root of the left lung, then descends within the thoracic cavity on the left side of the vertebral column, and continues into the abdominal cavity through the aortic hiatus in the diaphragm. Hence, it is described in several portions, namely, the ascending aorta, the arch of the aorta, and the descending aorta, which last is again divided into the (a) thoracic (above the diaphragm) and (b) abdominal aorta (below the diaphragm).

As with all other arteries, the aorta is made up of three layers: the *tunica intima* (composed mainly of endothelial cells), the *tunica media* (a muscular middle layer containing smooth muscle cells and elastic fibers), and the *tunica adventitia* (an outer layer of connective tissue). The aortic *tunica media* consists of smooth muscle and an extracellular matrix that is composed of ground substance, in which elastic fibers and collagen are embedded in a hydrated gel, allowing it to be a large elastic artery with inherent distensible properties.

Although the main function of the aorta is transporting oxygenated blood from the heart to the rest of the body, of equal importance is its capacity to distend and recoil in response to pulsatile flow, thereby reducing left ventricular afterload and facilitating diastolic perfusion of the coronary arteries [1]. The combination of smooth muscle cells (determining vasodilation and vasoconstriction), collagen (key for strength of the aortic wall), and elastin (permitting distension and recoil) within the walls of the aorta allows the aorta to maintain pulsatility and pressure across the entire circulation [2]. With advancing age, the aortic wall structure undergoes unfavorable changes, exacerbated by secondary effects due to adverse cardiovascular risk factors like systemic arterial hypertension, atherosclerosis, smoking, hypercholesterolemia, and diabetes, resulting in a decline in aortic elasticity and an increase in aortic circumference leading to aortic aneurysmal formation [2].

There is a growing body of data documenting progressive aortic dilation in patients with both repaired and unrepaired congenital cardiac defects [3, 4]. Several explanations might account for the increased incidence of aortopathy in adult congenital heart disease (ACHD), including a growing number of patients with congenital cardiac defects surviving into adulthood, along with improvements in cardiac imaging modalities, which provide unlimited and accurate assessment of aortic caliber, cardiac anatomy, and function in patients with congenital heart disease (CHD) [5].

As the life expectancy of CHD patients continues to improve, cardiologists will continue to be tested with the challenge of associated aortic sequelae like aneurysmal dilation, risk for aortic dissection, deteriorating aortic valvular disease, or progressive ventricular dysfunction. Although the natural history of aortic dilation (AD) has

been very well documented in certain genetic disorders, namely, Marfan syndrome [6], the expected course and functional significance of aortopathy in patients with structural forms of CHD remains less well defined [7].

A growing number of ACHD patients will go on to develop progressive AD and may be prone to associated complications including aortic valve insufficiency and aortic dissection or rupture; however, the risk of these complications in any individual patient with underlying CHD remains unknown. This chapter will review several congenital heart defects that are associated with aortopathies and will be divided into the following categories: (1) aortopathy associated with congenital BAV and coarctation of the aorta (CoA), (2) conotruncal lesions (TOF), transposition of the great arteries (TGA), double outlet right ventricle (DORV), interruption of the aorta, and truncus arteriosus (TA), and (3) patients with single-ventricle lesions after the Fontan palliation.

Bicuspid Aortic Valve

The congenitally bicuspid aortic valve (BAV) is the most common congenital malformation, occurring in approximately 2 % of the general population [8]. It is the most common cause of isolated valvular aortic stenosis in adults, with a male to female ratio of 3:1 [9]. Although the majority of cardiac events in BAV are due to valve dysfunction, there is a recognized increased prevalence of AD and aortic dissection (Fig. 146.1a–c). It is still unclear why patients with BAV may have a related aortopathy, aside from having specific associations with structural forms of CHD like CoA and an interrupted aortic arch, which has given rise to consideration of a common underlying aortopathy that may exist in patent with a BAV [10]. There is a five to tenfold increase in the incidence of aortic dissection in patients with BAV (when compared to patients with trileaflet aortic valves) without concomitant aortic stenosis, aortic coarctation, or systemic hypertension [11]. Warren et al. in children and Hahn et al. in adults showed that the aortic root was enlarged in patients with a BAV without aortic stenosis as compared to age- and sex-matched controls [12, 13].

It is also postulated that increased risk of aortic disease in patients with BAV (including normally functioning aortic valves) is mediated by coexisting defects in the aortic media by fragmentation of elastin, loss of smooth muscle cells, and increase in collagen [14, 15]. In acute aortic dissection, the underlying cause in approximately 15 % of patients was BAV, which is a higher proportion than those described with Marfan disease. In a study of 2,000 patients undergoing aortic valve surgery for BAV, 20 % required surgery for an ascending aortic aneurysm [16, 17]. These associations have led to the theory that congenital abnormalities of the aortic valve and the aorta may reflect a common developmental defect. The current understanding of the genetic basis of BAV has been improving and may be useful at preventing thoracic aortic complications in relatives of patients with known BAV. Patients with BAV can also have relatives with similar findings, thought to be transmitted as an autosomal dominant condition, with some affected relatives developing a thoracic aortic aneurysm despite the absence of a BAV [18]. With the current knowledge of the genetic basis of BAV, current guidelines recommend screening first-degree relatives of patients with BAV, or those with premature thoracic aortic disease (without significant known risk factors) to evaluate for the presence of a BAV [19]. It is also recommended that patients with BAV have both the aortic root and the ascending thoracic aorta evaluated for evidence of AD [16, 17, 20]. The ascending aorta should be imaged annually once the maximum diameter reaches 4.0 cm or more often depending on the progression of the aortic root [19]. Although the incidence of aortic dissection in BAV is nearly ninefold that of the general population, the overall incidence of aortic dissection in BAV is still low, given the large population with BAV.

Though still a subject of debate, surgery has been recommended to repair the aortic root or replace the ascending aorta in patients with BAV if the (a) diameter of the aortic root or

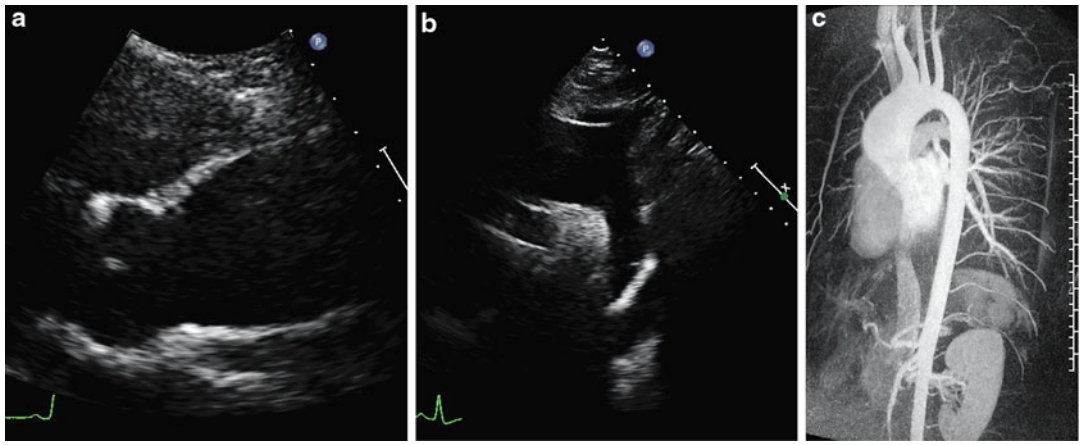

Fig. 146.1 A 20-year-old asymptomatic man with a BAV and an aneurysmal aortic root. (**a**) TTE PSLA view with aneurysmal dilation distal to the ST junction. (**b**) TTE suprasternal view with a dilated proximal transverse arch. (**c**) Cardiac magnetic resonance angiogram (MRA) showing the entire aorta. The maximum dimension of the ascending aorta was 5.1 cm

ascending aorta is greater than 5.0 cm, (b) if the rate of increase in diameter is 0.5 cm per year or more, or (c) if patients are undergoing aortic valve replacement (AVR) due to severe aortic stenosis or aortic regurgitation, with the diameter of the aortic root or ascending aorta being greater than 4.5 cm [21–23]. More recent studies have shown an acceptably low mortality rate for AVR when combined with ascending aorta repair in patients with BAV. Borger et al. found the 15-year freedom from aortic root complications (replacement, dissection) following AVR was 86 %, 81 %, and 43 % for aortic diameters of <40 mm, 41–44 mm, and >45 mm, respectively, at the time of AVR [23]. The decision of when to intervene with an elective surgery in an asymptomatic patient with a BAV and AD to prevent a potentially devastating dissection also depends on the patient's age, sex, and size, as well as other comorbidities and likelihood of surviving surgery. There is now a growing proportion of women reaching reproductive age with a BAV and associated AD. Such women with an aorta reaching 4.5 cm should be clearly counseled about the risks associated with pregnancy in the presence of AD [19].

With meticulous attention to best practice guidelines and close long-term follow-up with appropriate usage of imaging modalities to screen for aortic dilation and its complications, the long-term prognosis for patients with BAV is excellent and approaches an average life expectancy of 70 years [22].

Coarctation of the Aorta

Coarctation of the aorta (CoA) accounts for 5–10 % of all CHD and is associated with significant morbidity and mortality even after surgical repair. The prognosis of untreated coarctation is dismal, with 80 % dying prematurely from complications, including mortality from aortic dissection or rupture, heart failure, and intracranial hemorrhage [22]. Fortunately, most are treated in childhood and native CoA rarely presents in adulthood. Typically, the adult patient presents for routine follow-up after surgical palliation. Unfortunately, despite adequate surgical repairs, patients are at risk for several concerning long-term complications including systemic hypertension, re-coarctation (Fig. 146.2), aortic aneurysms (Figs. 146.3 and 146.4) and pseudoaneurysms, aortic dissection, sudden cardiac death, and patient-graft discrepancy from prior repairs [22]. A long-term follow-up study of patients repaired in childhood or adolescence demonstrated a significantly reduced

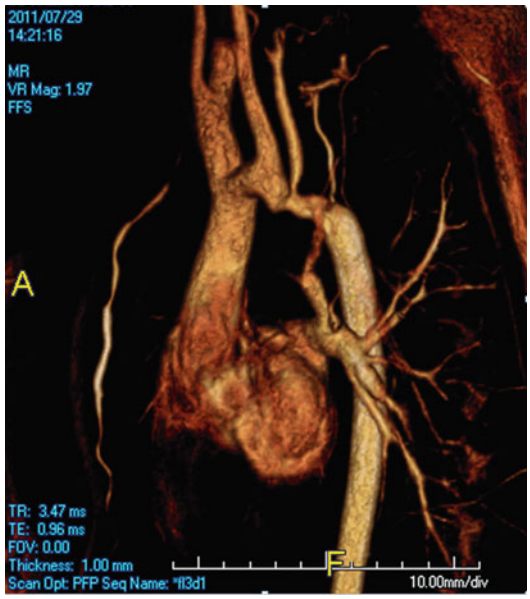

Fig. 146.2 A 50-year-old woman with CoA after surgical repair with a 12 mm Dacron tube graft at the level of the left subclavian artery. Three-dimensional volume-rendered MRA with evidence of re-coarctation just distal to left subclavian artery at the site of prior surgical repair

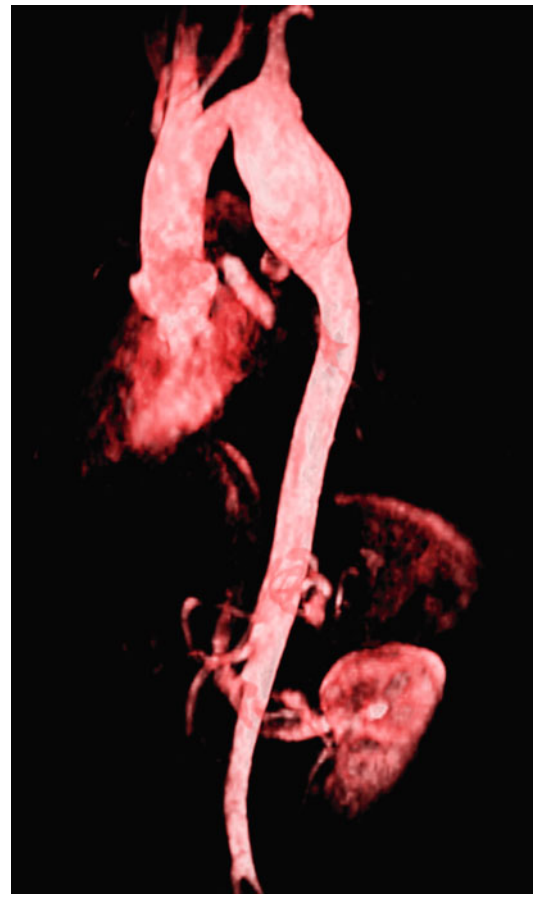

Fig. 146.3 A three-dimensional volume-rendered magnetic resonance angiogram (MRA) showing a large descending aortic aneurysm at the site of prior surgical repair in a 36-year-old woman with CoA. Maximum dimension was 4.8 cm with no evidence of aortic dissection noted

long-term survival with a mean age of death of approximately 38 years of age [24].

A variety of surgical options have been used over the past few decades and have continued to change over time. Treatments have included subclavian artery patch aortoplasty, patch aortoplasty, bypass of the coarctation segment with a graft, and tube-graft replacement. Endovascular techniques, such as balloon dilation and stent placement, including covered stent placement, have been used successfully and have become a less invasive alternative to open surgical procedures [25]. The type of palliation determines in large part the risks of re-coarctation and other complications.

An understanding of the underlying aortopathy in CoA will hopefully improve the long-term treatment and prognosis of patients with this pathology. Histologic abnormalities in the aortic wall with reduced elastic properties proximal and distal to the site of coarctation are evidence that CoA is a systemic vascular disease and not simply a narrowing of a discrete segment of the aorta. The presence of concomitant BAV in up to 85 % of coarctation patients and the strong histological similarity of aortic wall abnormalities between both entities are also suggestive of an inherited origin of aortic wall pathology [2].

Despite successful re-anastomosis of the aortic segments proximal and distal to the coarcted segment, long-term complications include persistent hypertension, re-coarctation, aortic dilation, aortic aneurysm, and aortic dissection or rupture [26]. Several factors have been identified as contributing to the development of hypertension, including the loss of aortic distensibility leading to a "functional" re-coarctation [27]. Patients with long-standing

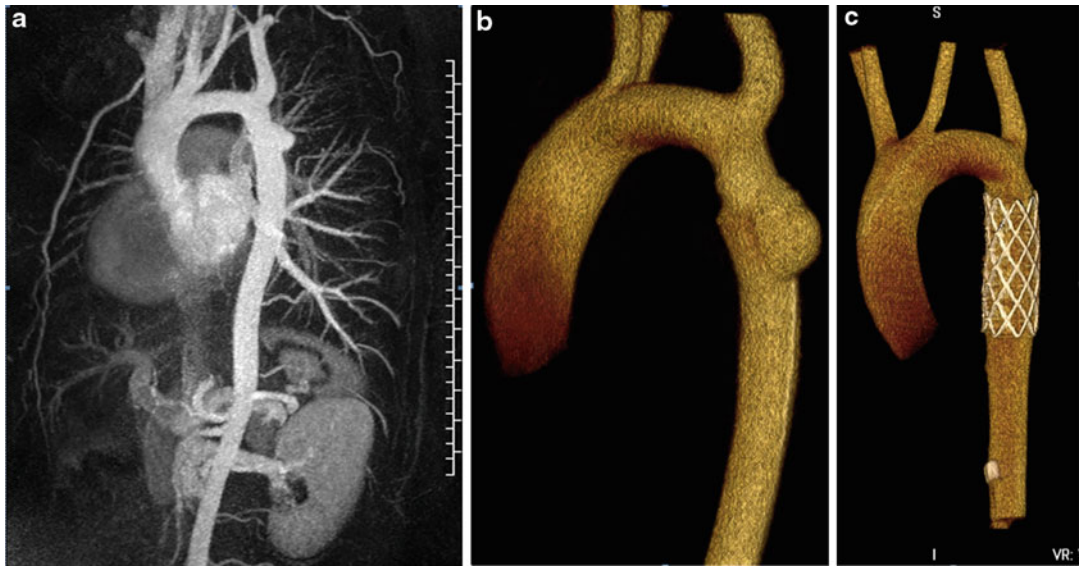

Fig. 146.4 A 26-year-old asymptomatic man with prior surgical repair of CoA during infancy. Routine screening three-dimensional reconstruction of a cardiac computed tomographic angiogram revealed a large saccular aneurysm at the site of prior repair with no evidence of aortic dissection

hypertension have been shown to have increased collagen in the aortic media and a decrease in smooth muscle [2]. The shape of the aortic arch may also predict adverse late events. A "Gothic" or triangular-shaped arch has been linked with increased vascular thickening, reduced aortic distensibility, and late-onset hypertension when compared to a "Romanesque" or more rounded arch [28]. In some patients, impaired LV function and increased LV mass have been found despite having coarctation repair and acceptable control of systemic hypertension, leading to the hypothesis that reduced elastic properties after coarctation repair can increase left ventricular afterload [29].

The appropriate type of treatment for native coarctation of the aorta in adults remains somewhat controversial. In particular, for women who are or will be of childbearing age after repair, there is a concern about the tissue integrity of the para-coarctation region, particularly during pregnancy. As such, caregivers may select direct surgical repair with excision of the para-coarctation tissue. For recurrent CoA (coarctation after surgical repair), catheter-based interventions (balloon or stent) are generally safe and the preferred alternative to surgery in the absence of confounding features (e.g., aneurysm or pseudoaneurysm formation, or significant coarctation that affects the adjoining arch arterial branches). For localized discrete narrowing, balloon angioplasty is an acceptable primary intervention but is still considered less suitable for long-segment or tortuous forms of coarctation. In many ACHD centers, surgery is reserved for patients who are unsuitable for or who have already failed percutaneous intervention [19].

Aortic aneurysms and rupture may occur years after successful repair of CoA [30]. This adverse late-term event appears to occur without recurrent coarctation and despite relief of systemic hypertension. For the majority of patients, aneurysm repair requires surgical intervention with resection of the aneurysm and graft placement. Unfortunately, there are no criteria to guide the timing of aortic aneurysm repair in this population. Pseudoaneurysms can also occur at the CoA repair site. They demonstrate an area of weakening with outpouching of the adventitial layer, usually along the suture lines, and are at a higher risk for rupture. Pseudoaneurysms should be considered for repair at the time of

initial diagnosis. Either surgical repair of the pseudoaneurysm or, in select cases, excluding the aneurysm with a covered stent percutaneously should be considered to remove the risk of rupture.

Though there is no apparent etiology for the higher incidence of hypertension and aortic dissection following coarctation repair, there is evidence of an intrinsic abnormality of the aorta that persists despite adequate repair [30, 31]. This "stiff" or less distensible aorta has also been described with essential hypertension, coronary artery disease, and Marfan syndrome and may be the underlying mechanism contributing to the late abnormalities associated with repaired coarctation of the aorta.

Adults with a history of successful coarctation repair should be meticulously followed for the presence of adverse sequelae such as re-coarctation, progressive AD, and dissection [26]. Routine screening with appropriate diagnostic modalities including echocardiography and at least one complete cardiac Magnetic Resonance Imaging (cMRI) or cardiac CT has been proposed to allow for early detection of these complications [32, 33].

Conotruncal Lesions

Conotruncal anomalies are a group of congenital heart defects that involve the outflow tracts of the heart and the great vessels. The outflow tract of the embryonic univentricular heart, also known as the conotruncus, begins as a common outlet but undergoes a highly choreographed sequence of events that results in the creation of separate left and right ventricular outflow tracts as well as the aorta and main pulmonary artery [34]. Of non-syndromic CHD, approximately 25–30 % are conotruncal cardiac defects [35]. Conotruncal cardiac defects include Tetralogy of Fallot (TOF), truncus arteriosus (TA), transposition of the great arteries (TGA), and double outlet right ventricle (DORV), and the very rare interruption of the aorta [36].

Although AD is commonly seen in conotruncal defects, such as TOF, d-TGA, and DORV, with some histological evidence of intrinsic aortopathy, there are at present no guidelines specific to conotruncal lesions regarding how to manage AD in these patients. Currently, aortopathies in patients with conotruncal lesions are managed similarly to patients with underlying connective tissue disease or a hereditary aortopathy.

Tetralogy of Fallot (TOF)

TOF is the most common form of cyanotic CHD and comprises 3.5–9 % of patients with CHD. The four components of TOF include the following: (1) malalignment ventricular septal defect (VSD), (2) overriding of the aorta, (3) right ventricular (RV) outflow tract obstruction/multilevel pulmonary stenosis, and (4) concomitant RV hypertrophy. The aortic arch is right-sided rather than left-sided in approximately 25 % of patients. Although TOF is the most commonly encountered cyanotic congenital heart defect in infancy, insidious development of progressive AD has been seen in adult survivors who have undergone surgical repair [37]. The underlying pathophysiology of ascending AD is unknown. In addition to previous long-standing volume overload, intrinsic histologic abnormalities in the aortic root and ascending aortic wall have been observed in several studies. Abnormalities of smooth muscles, elastic fibers, collagen, and ground substance in the tunica media of the ascending aorta were found to be prevalent in these patients, predisposing to aortic dilation, aneurysm, and/or aortic rupture [4, 37].

Single case reports of progressive AD first began appearing in the early 1970s [38]. It was not until the 1990s that the frequency of AD was described as occurring in up to 48 % of patients [39]. Since then, progressive aortic root dilation has frequently been described after TOF repair (15–88 %) [4, 40] (Fig. 146.5). Aortic dissections were first described in patients with repaired TOF in 2005 [41, 42].

It has been postulated that increased blood volume to the overriding aorta due to the additional volume through the VSD from the RV

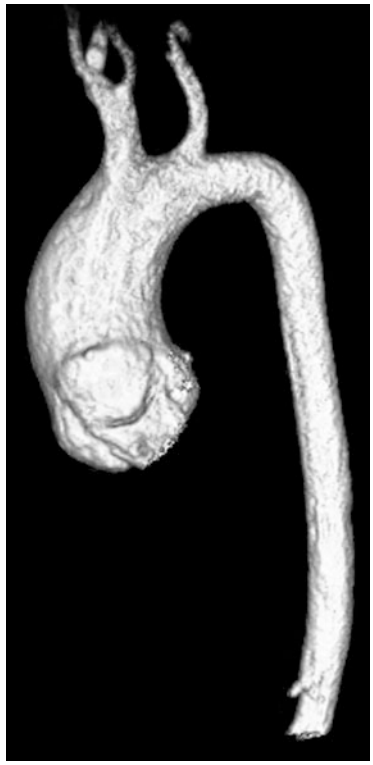

Fig. 146.5 A 52-year-old man with repaired TOF. Three-dimensional volume-rendered reconstruction of a cardiac MRA showing aneurysmal dilation of the ascending aorta with a maximum diameter of 5.2 cm

before surgical repair may result in increased stress on the aortic wall [4, 37]. Another hypothesis suggests that TOF itself may be a genetic aortopathy. Evidence supporting this hypothesis includes visualization of a dilated aorta on fetal echocardiography in TOF patients and multiple histologic studies using either pathologic specimens or surgical biopsies in infants with unrepaired TOF [2, 37]. Some studies have shown strong histological similarities of the aortic media as in Marfan syndrome in patients with repaired TOF with dilated aortas although a direct linkage to gene mutation(s) encoding for fibrillin-1 has not been clearly defined [2, 37]. Whether aortic wall pathology results from an intrinsic medial abnormality inherent to TOF itself or is secondary to the antecedent volume load through the aorta before repair (or perhaps a combination of the two) remains difficult to determine. Niwa et al. had investigated the ultrastructure of the great arteries in a series of patients who were already scheduled to undergo heart surgery. Of the 15 TOF patients in the series, all patients had at least grade 2 or 3 elastin fragmentation of the aorta [2]. Tan and colleagues found similar histologic evidence of an intrinsic aortopathy (including cystic medial necrosis, elastic fragmentation, and elastic lamellae disruption) in pathologic specimens of TOF which was present as early as a few days after birth, naïve to any surgical intervention [37]. Rutz et al. showed the incidence of AD and reduced aortic distensibility in patients with repaired TOF compared to age- and sex-matched controls [43]. The aortic diameters from the sinus to the level of the pulmonary artery bifurcation were larger, and the aortic distensibility was significantly reduced in patients with repaired TOF when compared to normal controls [43].

The ascending aorta should be imaged on a yearly basis in all patients with TOF if a progressive increase in aortic diameter, defined as an increase in aortic *z*-score, has been noted [4]. The risk of dissection in this patient cohort is being increasingly recognized; however, there remains no consensus guidelines in regards to surgical repair of a dilated aorta in these patients. In the only three cases reported in the literature, the absolute diameter of the aorta was $\geq$7 cm at the time of dissection, and the patients were much younger (18, 30, and 36 years, respectively) [41, 42]. Since all dissections occurred in patients with aortic diameters >5.5 cm, this raises the question as to whether ascending aortic replacement should be undertaken in those patients with an aortic diameter of $\geq$5.5 cm. Measurement of aortic stiffness, aortic curvature, and consideration of patient body size might enable us to further risk-stratify such patients [44, 45]. Prior studies have also reported the progressive nature of AD in TOF, which increased at a rate of 1.7 mm/year, in contrast to 0.03 mm/year in healthy controls [4]. Aortic dissection late after TOF repair in adults whose aortic roots exceeded 6 cm in diameter indicates that close monitoring of aortic dimensions is mandatory, especially when a dilated ascending aorta is present [37]. Elective surgical repair for the dilated aorta in

patients with repaired TOF has been actively debated with no current consensus guidelines. Aortic root surgery has also been considered in the case of progressive aortic valvular regurgitation and aortic root dilation exceeding 5.5 cm [7].

Given that TOF is the most common cyanotic congenital heart disease, and therefore one of the most common groups of ACHD patients, the need for more data to help determine ways to prevent aortic complications is growing daily. The potential for complications of AD that may necessitate surgical intervention is now increasingly recognized [37]. Despite overall excellent hemodynamic outcomes after surgery for TOF, there remains a concerning incidence of long term sequelae including aortic dilation. It is recommended that all ACHD patients with repaired TOF should have scheduled close and long-term follow-up with either echocardiographic and/or CMR evaluations as part of their routine follow-up to screen for early-onset complications [19].

Double Outlet Right Ventricle (DORV)

DORV is said to be present when both great arteries arise predominantly from the right ventricle with an incidence estimated at 127 per million live births [46]. DORV is not a single cardiac anomaly, but often used to describe an aberrant position of the great arteries in association with various cardiac anomalies in which the physiology is similar to that in VSD, TOF, TGA, or single ventricle (SV). DORV is defined as a form of ventriculo-arterial connection in which both great arteries arise completely or predominantly from the morphologic right ventricle. A VSD is almost always present; its location in relation to the semilunar valves may be subaortic (50 %), subpulmonary (30 %), uncommitted, or remote [47]. The relationship between the VSD and the great arteries, the relative outflow obstruction, and the anatomy of the atrioventricular valves determine the physiologic function, clinical course, surgical management, and long-term outcomes in DORV.

The ultimate goal of surgical management of DORV is to align the left ventricle with the systemic outflow and the right ventricle (RV) with the pulmonary outflow to achieve biventricular repair. Late complications after surgical repair of DORV vary with the individual anatomy and physiologic function, as well as the type of surgical procedure. The complications after repair of DORV with pulmonary outflow tract obstruction are similar to those seen after TOF repair, including chronic pulmonary regurgitation, dilation, and dysfunction of the RV, and arrhythmia. Aortic arch obstruction can be found in patients after coarctation or interrupted aortic arch repair. Those who undergo an arterial switch operation (ASO) may have the same problems described above for this procedure [48].

AD has been reported in adult patients with underlying DORV, but the data has been scarce (Fig. 146.6a, b). At the Mayo Clinic, from December 1973 through January 2008, 81 consecutive adults (median age, 34 years; range, 18–59 years) with conotruncal anomalies underwent operation on the aortic root, ascending aorta, or aortic valve. Primary cardiac diagnoses included TOF with or without pulmonary atresia in 60 patients, TA in 12, DORV in 6, and others in 3. Median ascending aortic size was 45 mm (23–80 mm). Operations included isolated aortic valve repair/replacement in 63 patients, combined AVR and reduction aortoplasty in 9, aortic root replacement in 7, and isolated ascending aortic replacement in 2. Median follow-up was 3.8 years (maximum 31 years). There were no ascending aortic reoperations after previous reduction aortoplasties or supracoronary ascending aortic grafts, and there were no late aortic dissections [49].

There is very limited data in regards to the incidence and rate of AD in patients with surgically repaired DORV. The true risk of either aortic dissection or rupture is currently unknown in this cohort of patients. Those who undergo an ASO may have similar complications, as described in the section on d-TGA s/p ASO including RVOT obstruction, coronary artery obstruction, and aortic root dilation [50, 51]. Currently there are no consensus guidelines to manage the aortic root

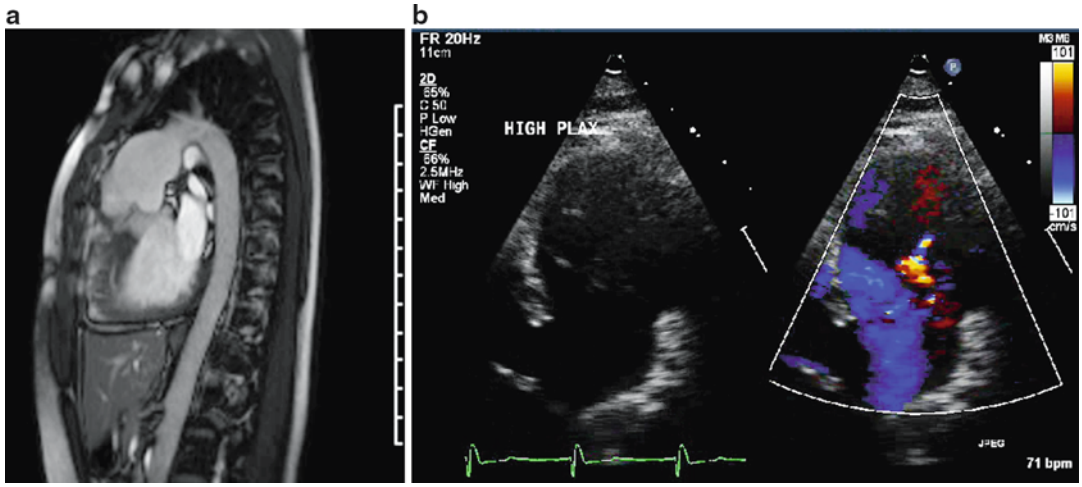

Fig. 146.6 A 22-year-old old woman with a history of DORV with a complex surgical history. (**a**) Cardiac MRI revealed a 5.1 cm ascending aortic aneurysm with no dissection. (**b**) TTE color Doppler images showing an eccentric jet of mild-to-moderate neo-aortic valvular insufficiency

in such patients with surgical repair. However, lifelong, systematic follow-up of their aortas to assess for AD, dissection, aneurysm, or pseudoaneurysm formation is strongly suggested.

Truncus Arteriosus

runcus arteriosus (TA) is an uncommon conotruncal anomaly with a reported incidence of 94 per million live births [46]. It is defined by the presence of a single artery arising from the heart with a single semilunar valve, giving rise to the coronary arteries, aorta, and at least one branch pulmonary artery. In the majority of cases, there is a sub-truncal VSD over which the truncal valve sits, similar to TOF. Associated cardiovascular and noncardiac anomalies are frequent [48]. Surgical repair usually follows the diagnosis. Typically, the VSD is closed with a patch so that the truncal valve is aligned with the LV (becoming the neo-aortic valve), and the pulmonary arteries are detached from the arterial trunk and connected to the RV with a valved homograft. Surgical repair of the truncal valve for stenosis or regurgitation is uncommon during the initial repair. The use of a nongrowing homograft in infancy makes additional operations inevitable as patients grow. Aortic arch interruption or coarctation is repaired at the same time.

The main complications after truncal repair are right ventricle–pulmonary artery conduit stenosis or regurgitation, branch pulmonary artery stenosis, neo-aortic valve (truncal) insufficiency or stenosis, VSD patch leak, and aortic arch obstruction. Most commonly, patients eventually require replacement of the right ventricle–pulmonary artery homograft because of increasing obstruction over time [52]. Long-term follow-up after TA repair performed in infancy has rarely been reported. A large retrospective review of surgeries since 1975 performed by Rajasinghe et al. assessed long-term outcomes among 165 patients who survived the initial hospital stay after complete repair of truncus arteriosus [53]. During the follow-up period, 107 patients underwent 133 conduit reoperations with a median time to conduit reoperation of 5.5 years. In addition, 26 patients underwent 30 truncal valve replacements. No patients had aortic root dilation or complications that needed surgical repair [53]. Large vessel (aortic and pulmonary arterial) dilation or dissection has rarely been reported in such patients, including young adults with unrepaired TA, highlighting the importance of the histological

abnormalities identified in the pulmonic trunk if placed under systemic pressure [54].

Overall, the 20-year survival and functional status appears to be satisfactory among patients who underwent complete repair of TA as infants, with conduit replacement or revision almost inevitably being the most common reason for repeat surgery in this group of patients.

Interruption of the Aorta

An interrupted aortic arch is defined by luminal discontinuity between the ascending and descending portions of the thoracic aorta. The discontinuity may be complete, or it may be spanned by an atretic fibrous band. Interrupted aortic arch is a rare condition; it is found in approximately 1 % of infants with a critical congenital heart defect [55]. An underlying genetic cause is thought to be responsible for many cases: A chromosome 22q11.2 deletion is identifiable in approximately 50 % of patients with an interrupted aortic arch, and 42 % of patients with DiGeorge syndrome have an interrupted aortic arch [55]. Surgical repair should be undertaken as soon as possible. In most institutions, the preferred surgical approach is direct anastomosis of the interrupted (or atretic) aortic segments. When the distance between the interrupted aortic arch segments is large, homograft augmentation may be added to the arch reconstruction. The use of a tubular conduit to bridge between the arch segments is usually reserved for unusually long-segment interruptions or for reoperations [48]. Long-term complications that require follow-up include residual or recurrent arch obstruction, aneurysm formation at the surgical site, residual obstruction of the LVOT, VSD patch leak, and left ventricular hypertrophy [47].

D-Transposition of the Great Arteries

D-TGA is defined by discordant connections between the ventricles and the great arteries. The aorta arises from the right ventricle, and the pulmonary artery arises from the left ventricle. The most common type of TGA is a d-loop (malposition) [47]. D-TGA is the second most common cyanotic congenital heart condition, with an incidence of 315 per million live births [46]. Associated defects with this lesion commonly include VSD, left ventricular outflow tract obstruction, and rarely CoA. Palliation of d-TGA became possible in 1950 with the development of the Blalock-Hanlon atrial septectomy, which improved inter-circulatory mixing. A physiologic repair using native atrial tissue was first reported by Senning in 1957, followed by Mustard's success with an atrial switch using a pericardial baffle in 1963. In 1975, Jatene [56] reported the first successful arterial switch operation (ASO), ushering in a new era of surgery for d-TGA. The ASO had become the predominant surgical strategy at most institutions by the late 1980s [57]. Patients with a large subaortic VSD and pulmonary stenosis may require a Rastelli procedure (valve conduit from right ventricle to pulmonary artery and patch created to tunnel blood from left ventricle to aorta) [58]. Today, the survival rate for infants with d-TGA is greater than 90 % with many patients surviving to adulthood with long-term outcomes often determined by the type of repair performed.

Transposition of the Great Arteries: Atrial Switch Palliation (Mustard/Senning)

Little has been written about the aorta in patients with d-TGA who have undergone a Mustard or Senning procedure. Given that morbidity and mortality is largely secondary to atrial or ventricular arrhythmias and congestive heart failure, evaluation of the aorta itself has not been an active area of investigation for most centers. Some of the research done so far has focused mainly on ventricular-arterial coupling as a mechanism of heart failure, exploring aortic distensibility and other measurements of aortic function rather than focusing exclusively on aortic diameter. Ladouceur and colleagues presented

results from 29 patients with d-TGA and intact ventricular septum, who had previously undergone atrial switch procedures [59]. Compared with age- and sex-matched controls, the patients had larger aortas at both the annulus (21.0 ± 3.6 vs. 17.6 ± 4.1 mm) and the sinus of Valsalva (30.0 ± 4.0 vs. 26.8 ± 4.2 mm) and also had lower aortic distensibility than controls (3.5 ± 1.6 vs. 5.3 ± 2.4 × 10^{-3} mmHg^{-1}). Rutz et al. showed that the incidence of aortic diameters from the sinus to the level of the pulmonary artery bifurcation was significantly larger in patients with d-TGA s/p atrial switch repair and the aortic distensibility was significantly reduced compared to age-matched controls [43].

There continues to be scarce data in regards to the incidence of AD in patients with d-TGA with atrial switch repairs. Although the follow-up studies of d-TGA after atrial repairs are important to the ACHD community, they are of less relevance to the d-TGA patient born in the current era now that d-TGA patients usually undergo the ASO. There are no consensus guidelines in regards to the imaging and management of the dilated aorta in patients with d-TGA after atrial baffle procedures. Echocardiographic imaging is recommended to evaluate the anatomy and hemodynamics with additional imaging with transesophageal echocardiography (TEE), cardiac CT, or cMRI used as appropriate, to evaluate the great arteries and veins [19].

Transposition of the Great Arteries: Arterial Switch Palliation

The transition from the atrial switch to the ASO spanned the decade of the 1980s. By 1985, patients with d-TGA were managed chiefly by ASO. Neo-aortic root dilation is common and may have significant implications in adulthood as afterload increases (Fig. 146.7a, b). In a report from Boston, freedom from aortic root dilation (z-score ≤3) was only 51 % at 10 years, and patients operated on more recently seem to be dilating earlier. However, dilation does not seem to be progressive once a z-score ≥3 is reached [60]. Thus, the functional fate of the neo-aortic root is unclear and may prove to be the most significant long-term issue in early adulthood [57].

Intrinsic aortic wall pathology in d-TGA has been postulated to be due to abnormal aortico-pulmonary septation or damage to the vasa vasorum and surgical manipulations during the ASO, thus predisposing the aorta to dilation, aneurysm formation, and even dissection [1]. Aortic distensibility may be reduced by impaired aortic elastogenesis and by scar formation at the site of anastomosis. High-grade medial abnormalities in the ascending aorta have already been observed during the neonatal period, suggesting that they are inherited analogously to Marfan syndrome. Aortic wall abnormalities may also develop due to structural wall differences between the two great arteries, as the former pulmonary arterial wall is exposed to higher systemic pressures after ASO, posing increased stress on the neo-aortic wall [2].

A high incidence of AR has been reported after ASO (30 % at 6 years after ASO) and is probably the result of a multifactorial process for which aortic root geometry, surgical techniques, and preoperative size discrepancy between the two great arteries are involved. In addition, AR appears to be functionally correlated with aortic root dilation and reduced elasticity of the proximal aorta, while the concomitant LV volume overload, increased LV dimensions and subsequent decreased LV ejection fraction [61]. Other authors have also reported that long-term follow-up has shown that some patients with ASO develop complications of the neo-aorta or neo-aortic valve requiring root or valve surgery [60, 62].

In order to help determine the natural history of neo-aortic dilation after ASO, Schwartz and colleagues evaluated a cohort of 335 patients who had undergone ASO for d-TGA either with or without a VSD, also including DORV with subpulmonary VSD [60]. Patients were followed for a median of 5.0 years. Overall, 33 % of the cohort had aortic dilation (defined as a neo-aortic root z-score ≥3.0 during at least one time point). When analyzing only the

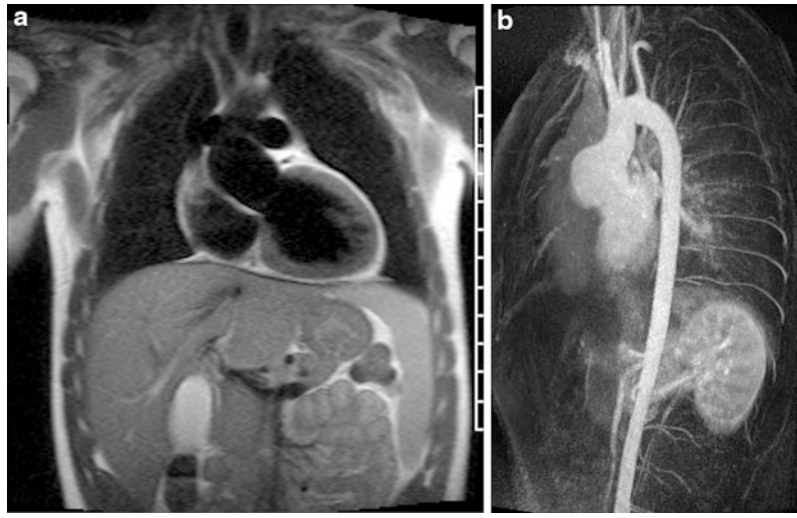

Fig. 146.7 A 16-year-old male with d-TGA after arterial switch operation presented with fatigue and exercise intolerance with a dilated ascending aorta measuring 5.0 cm. (**a**) Dark blood coronal image showing aneurysmal dilation of the sinuses of Valsalva and effacement of the sinotubular junction. (**b**) Maximum intensity projection of a cardiac MRA showing dilation at the level of the aortic sinuses and ascending aorta, with normal dimensions of the remainder of the aorta

patients with significant dilation, the mean z-score change was only minimal (increase of 0.05 per year), with a mean z-score of 4.6. Six patients had massive neo-aortic dilation with neo-aortic *z*-score $\geq$8.0, with three undergoing a neo-aortic root replacement. Although 49 % had AD at 10-year follow-up, only 5 % of the entire cohort required surgery on the neo-aortic root or valve. It should be highlighted that among those whose aortic root was dilated, the dilation tended to plateau rather than progress [60]. Though older patients with atrial baffle repair for d-TGA are at risk for arrhythmia, ventricular failure, and sudden death, it is thought that these sequelae are not likely to affect younger children with the newer arterial switch operation to the same degree. However, emerging complications, such as progressive coronary disease or progressive AD, have been described, and these may continue to arise as a result of this surgical approach [57].

Currently there are no agreement guidelines for ACHD patients with d-TGA after ASO in regards to management of the dilated aorta. Comprehensive echocardiographic imaging to evaluate the anatomy and hemodynamics in patients with d-TGA and prior ASO repair should be performed at least every 2 years. cMRI and CT angiography are recommended only periodically to evaluate the anatomy and hemodynamics in greater detail [19].

Congenitally Corrected Transposition (cc-TGA)

The second most common type of transposition of the great arteries is congenitally corrected TGA (cc-TGA), or l-loop transposition of the great arteries. cc-TGA is rare and is characterized by atrioventricular and ventriculo-arterial discordance. The condition is not cyanotic and may be discovered incidentally in asymptomatic patients. However, associated cardiac lesion such as a VSD, tricuspid valve abnormality (e.g., Ebstein anomaly), or pulmonary stenosis can coexist.

The minority of patients may be relatively normal from a functional standpoint, and survival to the seventh and eighth decades has been reported when no associated anomalies exist [63]. Complications include a progressive

incidence of complete AV block occurring at approximately 2 % per year, abnormalities of the systemic AV valve in up to 90 % of patients, and failure of the systemic ventricle. Systemic AV valve replacement is relatively common in the adult age group [64].

Very little has been written regarding dilation of the thoracic aorta in cc-TGA. A multicenter cross-sectional study done in 2000 showed an unexpectedly high prevalence of moderate or worse aortic regurgitation in 33 % of a population of 182 patients (mean age 32 ± 13 years, range 18–70 years) [65]. Unfortunately, no data on aortic root size was recorded in this study, so it is uncertain if this unexpectedly high prevalence of aortic regurgitation could be due to aortic dilation. It is difficult to gather comprehensive data on cc-TGA outcomes given how rare this diagnosis is in ACHD and how commonly there are other associated congenital anomalies [65]. Echocardiography-Doppler study and/or MRI should be performed yearly or at least every other year along with close clinical follow-up in adult patients with cc-TGA [19].

Single-Ventricle Physiology After Fontan Palliation

Many complex cardiac malformations are characterized by the existence of only one functional ventricle which has to maintain both systemic and the pulmonary circulations. The Fontan operation places the systemic and pulmonary circulation in series and is the treatment of choice for patients with a univentricular heart, resulting in near normalization of arterial saturation and removal of the chronic volume overload. As the oldest survivors of the Fontan palliation now enter their fourth decade of life, both the benefits and long-term sequelae associated with this palliation are increasingly being recognized. Several late complications including arrhythmias, heart failure, exercise intolerance, ventricular dysfunction, thromboembolic complications, hepatic dysfunction, protein-losing enteropathy, and worsening cyanosis have been described [66].

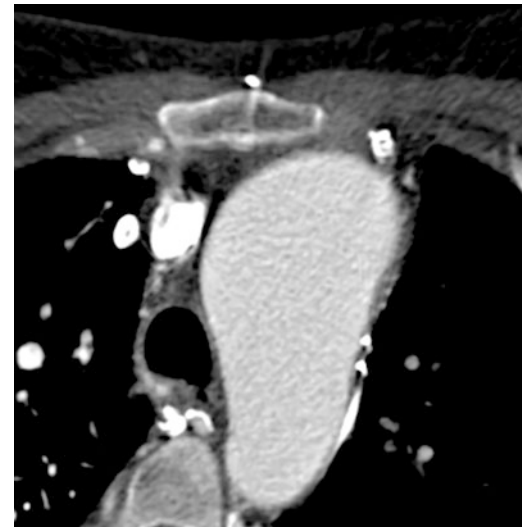

Fig. 146.8 A 38-year-old woman with DILV and malposed great arteries after Fontan palliation. Routine imaging revealed severe dilation of her ascending aorta extending to the mid-transverse aortic arch. Axial images from a CTA with maximum ascending aortic dimension of 5.2 cm with no evidence of aortic dissection

However, the role of aortopathy in older patients with SV physiology has not been well reported in the literature (Fig. 146.8). It is likely that aortic dilation will be encountered, since single-ventricle lesions often require reconstruction of the aortic arch in order to provide an adequate egress from the systemic ventricle to the systemic circulation.

The elastic properties of the reconstructed aorta are especially important in order to ensure efficient cardiac output in single-ventricle patients, especially in those with a single right ventricle, given that a morphologic right ventricle is more sensitive to increases in afterload than a morphologic left ventricle [67]. Hypoplastic left heart syndrome (HLHS) was the last common type of complex CHD to become amenable to csurgical therapy. It was not until 1983 that Norwood et al. reported successful palliation of a patient with HLHS to the Fontan circulation [68]. Thus, the longest possible follow-up for any patient with HLHS is just over 25 years, although additional predictions can be

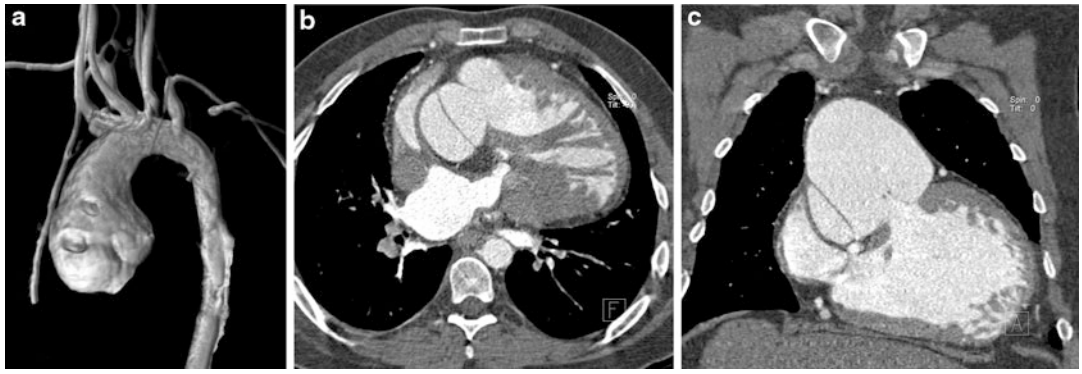

Fig. 146.9 A 27-year-old man with HLHS after Fontan palliation with a massive ascending aortic aneurysm. (**a**) Magnetic resonance angiography with three-dimensional volume-rendered reconstruction showing severe dilation of the aortic root. Maximum dimension was 8.2 cm. (**b**) Axial slices from a CTA 1 year later showed an acute aortic dissection, beginning distal to the neo-aortic valve. (**c**) Coronal CTA imaging showing the dilated ascending aorta and the dissection plane. There is a single systemic right ventricle that gives rise to the dilated aorta

extrapolated from data on late outcomes of the Fontan palliation for other forms of CHD [57]. The "neo-aorta" is often dilated especially after the Norwood palliation for several reasons (Fig. 146.9a–c). First, the pulmonary root, which is reconstructed into the neo-aortic root, is larger than the aortic root of healthy subjects [69]. Second, the ascending aorta and transverse aortic arch diameters are dependent on how much aortic patch material is used during the neo-aortic reconstruction. Prior studies have shown that patients with HLHS after Fontan palliation have a lower aortic distensibility index with elevated aortic stiffness [70].

Ultimately, the Fontan operation is a palliative procedure resulting in a unique single-ventricle physiology with a high incidence of long-term complications. The management of patients with SV physiology (especially HLHS) continues to evolve in the current era, including variations to the Stage I reconstruction using a right ventricle to pulmonary artery conduit (Sano operation), the choice between a lateral tunnel and extracardiac Fontan, aside from proposed interventional catheter techniques to replace one or more stages of the Fontan. These innovations promise that assessing long-term outcomes in adult patients with single ventricles will continue to require systematic long-term follow-up, with close analysis of the eventual neo-aorta with periodic echocardiography, cMRI, or CT evaluation [19, 57].

Conclusions

Aortopathy is common in several forms of CHD. Although AD and dissection are the most worrying complications, abnormal aortic elasticity has been reported and can result in impaired ventricular function. Multiple congenital heart defects including BAV, conotruncal defects, or patients with Fontan palliations have been associated with dilated aortas, dissection, aneurysm, or pseudoaneurysm formation in the past. Several of these defects are now considered part of a more generalized aortopathy with multiple prior studies documenting histologic evidence of elastin fragmentation, tying in to the hypothesis of an underlying tissue abnormality as a possible cause for the AD.

Patients with underlying BAV, CoA, repaired TOF, d-TGA, and Fontan physiology have been found to have AD. Long-term complications like aneurysmal dilation and dissections have also

been reported in such patients. Fifteen percent of all thoracic aortic dissections are associated with BAV. Rarely, aortic dissection has been reported in patients with repaired TOF, prompting increased awareness in regards to the dilated aorta in adults with CHD. However, there continues to be lack of data to determine when to intervene on a dilated aorta in conotruncal defects, such as TOF and d-TGA.

At this time, there are no exclusive guidelines for the management of thoracic aortic disease specific to patients with conotruncal defects or single ventricle physiology. Care should be individualized to each patient, with detailed assessment of the patient's underlying CHD, associated comorbidities, and the risk of surgery. Further research is necessary to determine adequate intervals for following progression of growth in aortic diameter and to determine the usefulness of novel imaging indices to further assess the dilated aorta in ACHD.

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