Calcification of the thoracic aorta is often associated with valvular and coronary calcification, reflecting an underlying atherosclerotic process. It has been found to be associated with an increased rate of mortality and cardiovascular disease. Porcelain aorta (PA) is extensive calcification of the ascending aorta or aortic arch that can be completely or near completely circumferential. This entity is rare in the general population, but it has an increasing incidence in older patients and in patients with coronary artery disease (CAD) or aortic stenosis (AS). The clinical relevance is based on the fact that it can complicate surgical aortic valve replacement (SAVR) for the treatment of severe AS by preventing safe access via the ascending aorta. PA is associated with increased morbidity and mortality, especially as a result of increased perioperative stroke risk. Recently, transcatheter aortic valve replacement (TAVR) has emerged as a less invasive and feasible treatment option in patients at high risk for conventional SAVR. In some series, ≥20% (5%-33%) of patients undergoing TAVR were diagnosed with PA. Inconsistencies in the definition and the use of different diagnostic modalities contribute to this wide range of PA prevalence. We reviewed the available published data to seek a consistent, clinically relevant definition based on contemporary imaging, a firm understanding of the pathogenesis and associations, and the clinical implications of this disease entity.

Definition and Diagnosis
PA has been used to address extensive circumferential or nearly circumferential calcification of the thoracic aorta such that it precludes safe cross-clamping or entry to the ascending aorta. However, there is no clear description or definition used; thus, cardiac surgeons and cardiologists use this term inconsistently. In the 1980s, Coselli and Crawford initially described 2 patients using the term PA for heavy calcification of the ascending aorta and aortic arch as diagnosed by chest radiography or palpation intraoperatively. They described replacing traditional aortic cross-clamping with femoral artery cannulation, profound hypothermia, and circulatory arrest with replacement of the ascending aorta to minimize the risk of aortic embolization or dissection. Svensson et al defined PA as calcification of the ascending aorta and aortic arch involving predominantly the aortic media. Leyh et al analyzed 1861 patients undergoing coronary artery bypass grafting (CABG) and found 23 patients (1.2%) with PA defined as circumferential severe calcification of the entire ascending aorta and proximal aortic arch. Several additional case reports and small case series of patients with PA undergoing cardiac surgery were published. The exact definition of PA varied between authors, but the common denominator that best describes the clinical problem is aortic calcification that interferes with aortic cannulation, aortic clamping, aortotomy, or central coronary bypass anastomosis, necessitating modification of the surgical technique to avoid complications.

Diverse modalities are used for the diagnosis of PA, and there is a lack of clear definition for how PA should be diagnosed. Not uncommonly, PA is recognized by manual palpation performed after sternotomy and exposure of the aorta at the time of cardiac surgery. Occasionally, a chest x-ray might reveal calcific outline of the ascending aorta or arch. Fluoroscopy during coronary angiography can also show diffuse, generalized calcification of the walls of the ascending aorta, suggesting the diagnosis of PA, but it is not an accurate modality for the assessment of the extent of aortic calcification. The most sensitive technique for detecting ascending aortic atheroma and calcification during open heart surgery is epiaortic echocardiographic scanning of the aorta in conjunction with manual palpation. Electron-beam computed tomography (EBCT) and multislice (spiral) CT are effective, noninvasive techniques for cardiac, coronary, and aortic calcification imaging preprocedurally. They are used to accurately evaluate the extent of ascending aorta and aortic arch calcification, thus differentiating between PA (circumferential) and less extensive aortic calcification, as well as determining the exact location in the ascending aorta and arch. CT is used more commonly for preoperative screening before aortic valve replacement both to exclude PA and to diagnose aneurysmal disease in younger patients. Examples of the various diagnostic presentations of a patient with PA are given in Figure 1.
Currently, in the TAVR era, the term PA is frequently used. It became an important factor for patient selection and sometimes serves as the primary indication for the TAVR approach even in intermediate-risk patients. The only randomized TAVR study, the PARTNER (Placement of Aortic Transcatheter Valves) trial, defined PA as nearly or completely circumferential calcification of the ascending aorta or aortic arch. Thus, extensively calcified aorta was found in 15.1% (54 of 358) of patients enrolled in the inoperable cohort of the PARTNER trial compared with 0.9% (6 of 699) of patients enrolled in the TAVR versus SAVR high-surgical-risk cohort. Rodés-Cabau et al reported TAVR procedures in 339 patients and defined PA as extensive circumferential calcification of the thoracic aorta as assessed by CT or fluoroscopy. PA was present in their cohort in 61 of 339 patients (18%).

Amorim et al recently suggested the use of the term PA when a circumferential calcification of the thoracic aorta is present at any given level. They proposed classification of PA into type I if circumferential calcification is present in the ascending aorta independently of further extension and type II if circumferential calcification is localized only in the aortic arch or descending aorta. Type I PA was further subdivided into type IA when there is no possibility to clamp the aorta during cardiac surgery and type IB when clamping is possible but at increased risk. This classification is helpful for guiding surgical relevance. Amorim et al also emphasized the importance of preprocedural chest CT to detect thoracic aortic calcification (TAC) and PA. The location of the circumferential calcification is crucial in surgical decision making. Although a narrow ring of calcium confined to the middle of the ascending aorta can safely be managed surgically without extensive technique modification, complete calcification of the aortic root or distal ascending aorta at the base of the innominate artery can mandate a much more extensive operation, necessitating aortic root and ascending aorta and partial arch replacement with reimplantation of the coronary arteries under circulatory arrest. This extensive surgery, which is frequently reoperative, is poorly tolerated in elderly, frail, and debilitated patients.

The nonuniformity in the definitions used to describe PA and the absence of this variable from conventional pre-TAVR risk scores (eg, logistic EuroSCORE or Society of Thoracic Surgeons score) motivated the Valve Academic Research Consortium to emphasize the importance of considering PA in the risk stratification performed by a dedicated heart team. The consortium defined PA or severely atherosclerotic aorta as “heavy circumferential calcification or severe atheromatous plaques of the entire ascending aorta extending to the arch such that aortic cross-clamping is not feasible.” Moreover, the consortium suggested noncontrast axial CT as the imaging tool of choice to evaluate calcification of the ascending thoracic aorta and aortic arch. Incorporation of CT and a standardized definition for the assessment of PA in the routine workup for all patients evaluated before cardiac surgery or TAVR may elucidate the true incidence and clinical implications of PA.

Figure 1. Diagnosis of porcelain aorta. A 67-year-old woman presented for evaluation of severe aortic stenosis. Her history included lymphoma treated with mediastinal radiotherapy at 35 years of age, hypertension, type 2 diabetes mellitus, chronic renal failure, ischemic heart disease, and coronary artery bypass graft surgery. A, Chest x-ray showing calcified ascending aorta and aortic arch (black arrows). B, Angiography during selective injection to the left internal mammary artery graft showing heavily calcified ascending aorta and aortic arch (black arrows). C, Transesophageal echocardiography showing calcification of the aortic valve and ascending aorta (white arrows). D and E, Chest computed tomography, oblique sagittal and axial views, demonstrating a nearly circumferential heavy calcification of the ascending aorta extending to the aortic arch.
Pathogenesis and Associations
The pathophysiological mechanisms contributing to the formation of a PA are not fully understood. Two independent processes lead to the formation of aortic calcification: atherosclerosis that occurs as a result of inflammatory response involving the tunica intima and calcification of mainly the medial layer of the aorta in the absence of atheroma.

Calcified PA involving predominantly the aortic media and atherosclerotic intimal plaque with calcifications might be 2 separate entities with considerable overlap.13

Atheromatous Aortic Disease: A Disease of the Tunica Intima
As part of the development of atherosclerotic plaques, calcium is deposited in the arterial wall by a process that is histologically similar to bone formation.23 Mineral deposits predominantly of apatite in the form of hydroxyapatite, carbonate apatite, and calcium deficient apatite may replace the accumulated remnants of dead cells and extracellular lipid, including entire lipid cores. Atherosclerotic lesions initially contain macrophage foam cells and fatty streaks.23,24 They may progress to intermediate and advanced lesions containing scattered collections of extracellular lipid droplets and lipid core, respectively (Figure 2A). When the lipid core and other parts of the lesion become calcified, it may be referred to as type Vb lesion.23

Several mechanisms contribute to the intimal calcification process: Apoptotic cell death of vascular smooth muscle cells (VSMCs) and macrophages can induce the generation of extracellular matrix vesicles that may serve as sites for calcification and can induce increased local concentration of calcium and phosphate ions.25 Inflammation in a plaque may also elevate extracellular ion concentration.26 Elastin can act as a calcifying protein that contributes to the calcification process.23 VSMCs and intimal macrophages produce many calcification-regulating proteins commonly found in bone such as osteopontin, osteocalcin, osteonectin, matrix Gla protein, alkaline phosphatase, and bone morphogenetic protein 2 (BMP-2).

Figure 2. Mechanisms of thoracic aortic calcification: intimal atherosclerotic calcification versus medial nonatherosclerotic calcification. A, Intimal calcification. Endothelial injury initiates the creation of macrophage foam cells and the migration and proliferation of vascular smooth muscle cells (VSMCs) from the media to the intima. Layers of foam cells and lipid-laden VSMCs are designated as fatty streaks. Intermediate lesions contain scattered collections of extracellular (EC) lipid droplets. Advanced lesions are characterized by larger, confluent lipid core. As this process progresses, a prominent fibrous connective tissue (CT) can be formed, followed by intimal calcification. Several mechanisms contribute to this process: Elastin can act as a calcifying protein; apoptotic cell death of VSMCs and macrophages can induce extracellular matrix (ECM) vesicles generation that may serve as sites for calcification; and inflammation may elevate extracellular calcium and phosphate concentration. Inflammation in a plaque may also elevate extracellular ion concentration. Elastin can act as a calcifying protein that contributes to the calcification process. VSMCs and intimal macrophages produce many calcification-regulating proteins commonly found in bone such as osteopontin, osteocalcin, osteonectin, matrix Gla protein (MGP), alkaline phosphatase, and bone morphogenetic protein 2 (BMP-2). B, Medial calcification. Uremia, radiation, and vascular inflammation induce a phenotypic change of VSMCs into osteoblasts. Transformed osteoblasts produce a number of bone-associated proteins not normally expressed in the vessel wall, including alkaline phosphatase, bone sialoprotein, bone Gla protein, and BMP-2. There is also evidence of downregulation of MGP, which is known to inhibit medial calcification. Apoptotic bodies derived from dying VSMCs and matrix vesicles released from VSMCs are also essential contributors. Elastin degradation, mediated by matrix metalloproteases (MMPs), serves as an initiating nidus for medial calcification. As this process progresses, it can eventually form a dense circumferential sheet of calcium crystal in the center of the media, bound on both sides by VSMCs and often containing bone trabeculae and osteocytes.
2, 23, 25, 27, 28 VSMCs therefore appear to adopt an osteogenic phenotype (Figure 2A).

TAC was found to be associated with the conventional cardiovascular risk factors, that is, aging, hypertension, smoking, dyslipidemia, and diabetes mellitus29–31 (Table). It was also found to be related to coronary artery calcification30 and increased risk of cardiovascular events and mortality.1, 32 Kälsch et al30 assessed the EBCT scans of 4025 participants and increased risk of cardiovascular events and mortality.1, 32 Kälsch et al did not present subanalyses of AAC versus DAC. The Multi-Ethnic Study of Atherosclerosis (MESA) trial evaluated thoracic aortic wall calcification using multidetector CT scans of 6814 participants without known cardiovascular disease.29 The prevalence of calcification in this cohort was lower: 56 participants (1%) had isolated AAC, 1675 (25%) had isolated DAC, and 178 (3%) had both. Current smoking, hypertension, dyslipidemia, and aging were found to be associated with AAC. Different CT scan acquisition methods for calcification evaluation and the diverse populations observed in these trials might explain the different prevalences of TAC.

Nonatheromatous Aortic Disease: A Disease of the Tunica Media
Thoracic AAC in its extreme form, PA, does not necessarily share the exact pathophysiological mechanisms with atherosclerosis33. Vascular medial calcification occurs independently of intimal calcification and atherosclerosis.33 Uremia, radiation, or vascular inflammation induces a phenotypic change of VSMCs into osteoblasts.24 Transformed osteoblasts in the media produce a number of bone-associated proteins not normally expressed in the vessel wall, including alkaline phosphatase, bone sialoprotein, bone Gla protein, and bone morphogenetic protein 2.24, 25, 28 There is also evidence of downregulation of matrix Gla protein, which is known to inhibit medial calcification.25 Apoptotic bodies derived from dying VSMCs and matrix vesicles released from VSMCs are essential contributors to medial calcification.24, 35 They enable nucleation of mineral crystals by the matrix proteins and concentration of calcium and phosphate in preparation for mineralization.24 Another contributing factor is elastin, which serves as an initiating nidus for medial calcification. Elastin degradation is mediated by matrix metalloproteases. This process has been associated with the development of arterial medial calcification.24 Moreover, it is possible that elastic lamellae create a physical barrier between VSMCs, preventing their migration and limiting calcification to the media layer.3 As this process progresses, it can eventually form a dense circumferential sheet of calcium crystal in the center of the media, bound on both sides by VSMCs and often containing bone trabeculae and osteocytes (Figure 2B). Medial calcification typically occurs in lower-limb arteries such as the femoral artery, but it is also a common finding in the aorta.36 Aging is associated with a number of structural and functional changes of the arterial wall media, including hypertrophy, extracellular matrix accumulation, and calcium deposits.37, 38 It evokes paracrine osteogenic signals that promote aortic calcium deposition. There is a significant acceleration of this process in patients with metabolic disorders such as the metabolic syndrome or diabetes mellitus39, 40 (Table).

The differentiation between intimal atherosclerotic calcification and medial nonatherosclerotic calcification has potentially important clinical implications in the surgical and percutaneous management of valvular and CAD in patients with PA. The presence of a heavily calcified atheromatous aorta is associated with a significantly increased risk of embolic stroke and peripheral embolism during conventional cardiac surgery.6, 15, 42 On the other hand, when calcification of the aorta is limited to the tunica media, it excludes a significant source of embolization because the intima is relatively intact without exophytic lesions but precludes safe cross-clamping or cannulation of the aorta. Nonetheless, the currently available diagnostic methods used to detect TAC, including CT, do not discriminate intimal from medial calcification, thus precluding a clear, clinically valuable separation between patients with PA.1, 42

Special Disease Groups Associated With PA
Chronic Kidney Disease
Patients with chronic kidney disease (CKD) demonstrate both accelerated calcification of atherosclerotic plaques and diffuse calcification of the media of large arteries (Monckeberg sclerosis).25, 27 CKD often results from hypertension and diabetes mellitus, and patients with CKD are among the highest-risk group for cardiovascular morbidity and mortality. Intimal atherosclerotic calcification is a common pathological process among patients with CKD with multiple cardiovascular risk factors.24 A second and distinct mechanism of calcification in patients with CKD is medial calcification, a process of metabolite-induced (toxic) vascular changes in the absence of lipid...
deposits. As this process progresses, it can lead to increased vascular stiffness and progressive loss of the cushioning function of blood vessels (arteriosclerosis). CKD-related risks such as phosphate retention, increasing serum calcium, hyperparathyroidism, vitamin D deficiency, and duration of dialysis contribute to this type of calcification. DeLoach et al used EBCT scans of 112 renal transplant recipients to evaluate AAC and DAC. Aortic calcification was found in 34% of patients and was associated with an increased incidence of cardiovascular events and higher mortality. Excessive vascular stiffness of the aorta may lead to congestive heart failure, left ventricular hypertrophy, elevated pulse pressure, coronary ischemia, or sudden death resulting from arrhythmia. These are all possible contributors to the higher cardiovascular morbidity and mortality in patients with CKD.

Radiation-Induced Cardiovascular Disease
Mediastinal radiation may also cause both accelerated atherosclerotic calcification of the aorta and dystrophic calcification, which has a sharp, pencil-like outline as a sequel of scarred aortic intima or media. Radiation-induced heart disease encompasses a range of deleterious effects on the heart, including aggressive CAD, valvular disease, constrictive calcified pericarditis, advanced diastolic dysfunction, and conduction abnormalities. The cumulative incidence of radiation-induced heart disease is between 10% and 30% by 5 to 10 years after treatment. Desai et al studied multidetector CT angiography studies of 117 patients with radiation-induced cardiac disease. AAC was noted in 69 patients (60%), with 18 patients (15%) having a moderate calcification and 15 severe calcification was 8.8% for men and 19.1% for women. Increased thoracic and abdominal aortic calcification.62 Possible explanation for female predominance in aortic calcification might be that, as mentioned above, aortic calcification represents not only the atherosclerotic process but also bone calcium metabolism. Redistribution of calcium in elderly populations also revealed inconsistencies in sex distribution, showing a male predominance in some and a female predominance in others. Nasir et al directly evaluated sex differences in TAC. TAC was present in 8549 asymptomatic individuals (69% men; mean age, 52 years) revealed higher prevalence of TAC in women compared with men (25% versus 21%; P<0.0001). Adjustment for traditional CAD risk factors did not change these results. A possible explanation for female predominance in aortic calcification might be that, as mentioned above, aortic calcification represents not only the atherosclerotic process but also bone calcium metabolism. Redistribution of calcium in elderly women during the development of osteopenia or osteoporosis from bone to soft tissue, including the aorta, results in increased thoracic and abdominal aortic calcification.

Clinical Implications
PA is usually an incidental finding in patients being evaluated for cardiovascular or pulmonary diseases. The presence of a heavily calcified ascending aorta or arch is asymptomatic, a fact that precludes true evaluation of the prevalence of PA in the general population. Inconsistencies in definitions and the use of different diagnosis modalities, as mentioned above, also restrict the ability to assess the true prevalence of PA. CT scan assessments of AAC without definition of severity revealed calcification in 2.7% to 42.9% of patients without known cardiovascular disease. Hunold et al found severe calcification of the thoracic aorta in 13 of 1812 patients (0.7%) with known or suspected CAD who underwent an EBCT scan. The prevalence of PA was found to be 7.5% in patients evaluated for AS, between 1.2% and 13.6% in patients undergoing valvular or coronary revascularization surgeries, and between 5% and 33% in the patients undergoing TAVR.

PA and Cardiovascular Risk
As discussed earlier, TAC and PA have been associated with cardiovascular risk factors, as well as with CKD and radiation exposure. There is growing evidence that identification of ascending aortic or aortic arch calcification can also be related
independently to a higher risk of cardiovascular events and mortality. The largest study to examine the relation between TAC and mortality evaluated a cohort of 8401 asymptomatic individuals who underwent an EBCT scan for the assessment of an underlying coronary heart disease risk. 69 Multivariable analysis revealed that the presence of aortic calcification was independently related to increased mortality during an average follow-up of 5 years (hazard ratio, 1.78; P=0.002). However, this study did not discriminate between regions of TAC. Vehmas70 examined the spiral CT of 504 men screened for lung cancer. Participants who had aortic arch calcification were found to have increased mortality independently of covariates. Both AAC and DAC were also found to be associated with increased mortality, although this association was weaker.

Eisen et al1 evaluated TAC in 361 patients with stable angina pectoris who had a spiral CT and were followed up for a period of 4.5 to 6 years. TAC was found in 253 patients (70%): 41 patients (11.4%) had isolated AAC, 79 patients (21.9%) had isolated DAC, and 133 patients (36.8%) had calcification in both the ascending and descending aorta. All 19 patients who died during follow-up had TAC. Adverse cardiovascular events were also higher among patients with TAC. The study of Gondrie et al71 demonstrated that calcifications in all 3 parts of the thoracic aorta diagnosed in chest CT scans performed for noncardiovascular indications in 1164 patients were separately and independently related to fatal and nonfatal cardiovascular disease events during a mean follow-up of 17 months. Jacobs et al72 examined the influence of severe AAC or DAC found on chest CT of 1723 patients on cardiovascular disease events. The risk of any cardiovascular disease event during a mean follow-up of 18 months was increased by a factor of 2.7 among patients with severe TAC.

In comparisons of AAC and DAC with regard to increased stroke risk, there have been contradictory findings. Jacobs et al72 found increased risk of stroke only in patients with AAC, whereas Tanne et al73 found increased risk of stroke only in patients with DAC. An association between cerebrovascular events and calcification of the descending rather than the ascending aorta may suggest that calcification of the thoracic aorta is not a direct causative factor for embolic stroke but rather a marker of increased burden of atherosclerotic disease. X-ray or CT assessment of the thoracic aorta may show only the calcified lesion and not the softer, atheromatous part of the plaque that is presumed to be more vulnerable. Cohen et al74 performed an ascending aorta and aortic arch transesophageal echocardiographic evaluation of 334 patients admitted with stroke. They found that the risk of subsequent cerebrovascular or other embolic events was systematically higher during a mean follow-up of 2.3 years in patients with thicker aortic plaques and in patients who had plaques without calcification. However, in several studies, aortic arch calcification diagnosed with chest x-ray or CT was found independently to be related to increased rate of cerebrovascular events. 62,75 supporting the theory that TAC and its extreme form, PA, are indicators of atherosclerotic burden in populations with multiple cardiovascular risk factors. Furthermore, there is increased evidence of an association between AAC and CAD. Histopathological and clinical studies have shown a close correlation between coronary artery calcification and the existence and severity of CAD. 76,77 Both coronary calcium and AAC can be diagnosed noninvasively by spiral CT or EBCT. AAC was found to be independently related to coronary artery calcification, 30,78 to multivessel coronary disease in women, 2 and to significant coronary stenosis requiring revascularization.79

**PA and Cardiac Surgery**

An important clinical implication of PA is an increased risk of stroke during cardiac surgery caused by embolization of atheromatous material resulting from manipulation of the ascending aorta. 14 It is well established that atherosclerosis of the ascending aorta diagnosed by palpation or epiaortic ultrasound during cardiac surgery is an independent predictor of short- and long-term neurological events and mortality. 66,80 van der Linden et al81 examined the relation between atherosclerotic disease in the ascending aorta and postoperative stroke in 921 patients who underwent cardiac surgery. Atherosclerotic disease in the ascending aorta was detected in 26.2% of patients, and calcification was noted in approximately half of these patients. The incidence of postoperative stroke was 1.8% in patients without and 8.7% in patients with atherosclerotic ascending aorta disease (P<0.001). Extensive involvement by the disease of more than half of the ascending aorta increased the risk of stroke to 33%. Interestingly, the presence or absence of calcification in the ascending aorta was not a predictive risk factor for stroke, indicating that medial disease is not prone to embolization as easily as soft, noncalcified atherosclerotic disease. However, the presence of heavily calcified PA is associated with a significantly increased risk of cerebral embolism during cardiac surgery. 6,41 Severe AAC interferes with aortic cannulation, aortic clamping, aortotomy, and central coronary bypass anastomosis, necessitating modification of the surgical technique to avoid complications such as aortic dissection, surgically unreconstructable ascending aorta, or release of thromboembolic material that may cause periprocedural stroke. 2,15,82

Several modifications have been used in isolated CABG procedures to avoid cannulation and clamping of the diseased ascending aorta. The most common modification is a “no touch” technique that totally avoids manipulation of the ascending aorta. This is most readily accomplished by off-pump techniques and all arterial grafting using bilateral internal mammary grafts with the addition of a radial artery as a side Y or T graft. Alternatively, if the aorta is not totally calcified and a noncalcified area is ascertained by epiaortic scanning and palpation, a “clampless” proximal anastomosis can be performed using a Heartstring device. 3,59,60 Lev-Ran et al59 retrospectively compared the results of coronary revascularization between cardiopulmonary bypass with femoral artery cannulation in 15 patients and off-pump CABG in 41 patients with PA. There was only 1 case of perioperative mortality (2.4%) and there were no cases of perioperative stroke or transient ischemic attack in the off-pump CABG group. The main disadvantage of this strategy in their report was incomplete revascularization in 24.3% of the patients. The cardiopulmonary bypass group had 1 mortality (6.6%), 3 perioperative strokes or transient ischemic attacks (20%), and a lower rate of incomplete revascularization (6.6%). Other options include
femoral or axillary artery cannulation for cardiopulmonary bypass, intraluminal balloon catheter as a substitute for external clamps, avoidance of proximal graft anastomoses on the ascending aorta, and CABG performed during deep hypothermic circulatory arrest. Extended procedures such as ascending aorta endarterectomy, patch aortoplasty, and graft replacement of the ascending aorta have also been described.

Mitrval valve procedures can also be performed without aortic cross-clamping by hypothermia and a fibrillating heart. Tricuspid valve and other right-sided procedures can be performed on a beating heart without the need for aortic cross-clamping or manipulation. Aortic valve procedures in patients with severe calcification of the ascending aorta mandate the greatest modification of usual techniques. Techniques for SAVR in patients with PA include SAVR under deep hypothermic circulatory arrest. With this technique, the ascending aorta can be managed with several strategies: aortotomy low on the ascending aorta if any noncalcified area is present or replacement of the ascending aorta and ascending aortic endarterectomy. Although most of the procedures can be performed with <20 minutes of circulatory arrest, there may be some reluctance to perform deep hypothermic circulatory arrest in the setting of advanced age and significant comorbidities. Another option to avoid aortic cross-clamping and manipulation in patients with severe AS is the apico-aortic conduit technique. It is an aortic valve bypass surgery that uses a conduit containing a prosthetic valve that relieves AS in case of a concomitant calcified descending aorta, which is usually present.

Each of the methods described above for SAVR in patients with severe AS and PA is technically challenging, requires significant surgeon experience, and does not completely avoid manipulation of the heavily calcified ascending aorta. Therefore, no standard treatment has emerged, with surgical management individualized to the particular patient and to the location and extent of the calcification present. There is still a considerable risk of perioperative mortality or stroke among these patients. Interestingly, Nishi et al evaluated preoperative CT scans of 11 patients with PA and observed that the calcification grade just below the innominate artery is significantly lower than that at the level of the pulmonary artery, where aortic cross-clamping is usually applied. Calcification in <75% of the circumference was considered sufficient to allow soft cross-clamp of the aorta just below the innominate artery in all 11 patients, and SAVR under cardiopulmonary bypass with mild hypothermia was completed without peri-procedural mortality or stroke.

PA and TAVR

With the evolution of TA VR, a new technology to treat high surgical risk patients with severe AS that avoids aortic cross-clamping and cardiopulmonary bypass has become available. A few studies have directly evaluated and reported the impact of PA on the outcome of patients after TAVR. Rodés-Cabau et al were the first to report the impact of PA defined as extensive circumferential calcification of the thoracic aorta as assessed by CT or fluoroscopy. In a Canadian registry of 339 patients who underwent balloon-expandable TAVR, 61 patients (18%) had PA. Patients with PA in this study were younger, more frequently female (75.4% versus 50.7%), and exhibited a lower Society of Thoracic Surgeons score. The procedure was successful in 98.4% of the patients with PA, but valve malposition requiring implantation of a second valve tended to be more frequent in these patients (6.6% of patients with PA versus 1.8% in patients without PA; P=0.059). The stroke and 30-day mortality rates were 1.6% and 11.5%, respectively, with no difference in patients without PA. Patients with PA tended to have a better survival rate at the 1-year follow-up (86% versus 74%; P=0.14).

Pascual et al reported the results of CoreValve implantation in 449 patients using the transfemoral or transaxillary approach. Thirty-six patients (8%) had PA that was diagnosed with fluoroscopy. The procedural success rate was high (97.3%), with no differences in the percentage of malapposition or any other periprocedural complication between patients with and without PA. There was no significant difference in 2-year survival between the 2 groups. In a relatively large German registry, Zahn et al reported the results of TAVR in 147 patients (10.7%) with PA compared with 1227 patients without PA. Either CoreValve or Edwards valves were used with transfemoral, transsubclavian, transaortic, or transapical approaches. The procedural success rate was high (97.3%) in both groups. Coronary ischemia complicated the procedure more often in patients with PA (2.1% versus 0.1%), and there was a trend toward a greater 30-day stroke rate (5.5% versus 2.8%; P=0.08). The 30-day mortality was higher among patients with PA (12.9% versus 7.6%; P=0.03), but multivariate analysis did not show that PA was an independent predictor of inhospital death or stroke. The main limitations of this study were that the diagnosis of PA was left to the treating physician without uniform criteria or diagnostic modality and that the observed range in prevalence of PA between participating hospitals was 0% to 70%.

Furthermore, a recent subanalysis of the inoperable arm of the PARTNER trial performed by Makkar et al found PA to be the commonest reason for technical inoperability (39 of 85 patients, 46%). PA and other reasons for technical inoperability in this study (eg, mediastinal radiation or chest deformities) were associated with similar procedural outcomes and better 2-year outcome than seen in patients who were excluded from surgery for clinical reasons. The comparative data presented above suggest that, unlike in SAVR, PA does not mandate significant procedural modifications and does not affect procedural outcome in TAVR. Therefore, TAVR is expected to become the standard of care for patients with severe AS and PA. PA is a particular subset of “technically inoperable” patients who are particularly well suited for TAVR because there are usually no other significant comorbidities present and the surgical alternatives are usually more extensive procedures than isolated SAVR.
Conclusions

PA is an extensive circumferential or nearly circumferential calcification of the ascending aorta extending to the aortic arch. Chest CT is the most effective method to diagnose calcification of the thoracic aorta and PA. Two independent processes lead to the formation of aortic calcification: atherosclerosis involving the intima and calcification of the medial layer in the absence of atheroma. These processes have separate but overlapping pathophysiology and clinical associations. There is growing evidence that the identification of ascending aortic or aortic arch calcification can also be related independently to a higher risk of cardiovascular events and mortality. Patients with PA undergoing conventional cardiac surgery require procedural modifications to attenuate neuroembolic risk and safe aortic entry and closure. TAVR is a safe and efficient option for patients with severe symptomatic AS and PA. Routine use of chest CT for the assessment of PA in the workup of all patients evaluated before cardiac surgery or TAVR may help elucidate the true incidence and clinical implications of PA.

Disclosures


**KEY WORDS:** aorta □ aortic diseases □ calcification, physiologic □ cardiovascular diseases
Porcelain Aorta: A Comprehensive Review
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In the article by Abramowitz et al, “Porcelain Aorta: A Comprehensive Review”, which was published in the March 3, 2015 issue (Circulation. 2015;131:827–836), Figure 2 was incorrect. The correct version of Figure 2 is provided below.

The corrections have been made to the current online version of the article, which is available at http://circ.ahajournals.org/content/131/9/827.full.