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Letter to the Editor

Transcatheter aortic valve replacement will be standard of treatment for severe aortic stenosis with porcelain aorta

Calcification involving thoracic aorta is a known pathology and it is often associated with calcification extending to valvular, coronary and carotid arterial system.[1] It reflects a common atherosclerotic calcific pathology affecting the cardiovascular system.[2] Porcelain aorta (PA) is the term used to refer to the condition characterized by extensive calcification of thoracic aorta.[3] Concomitant severe aortic stenosis (AS) or severe coronary artery disease (CAD), which often compound PA, significantly increase cardiovascular mortality and hence this condition needs to be addressed meticulously. Aortic valve replacement (AVR) in the presence of PA is very complex surgical procedure associated with increased complications and mortality but transcatheter AVR (TAVR) today has eased out this unmet clinical need. Inoperability for AVR in PA is an example where surgery is challenging because of technicalities rather than the high clinical risk from several other comorbid conditions seen in these patients.[4,5]

PA has been defined as a structural disease of the aortic wall where extensive and circumferential calcium deposition occurs in the thoracic aorta that can be detected by computed tomography (CT) or fluoroscopy. Calcium is present circumferentially or partly in the ascending aorta, the arch and the descending thoracic aorta. The calcium deposition may occur in the intima alone, as seen in those with atherosclerotic pathology. People with hypertension, diabetes, dyslipidemia and smoking generally have such intimal calcification and it usually affects the very elderly population. On the other hand, a non-atheromatous calcium deposition in the thoracic aortic media can also occur leading to the genesis of PA. This form of aortic calcification is more commonly seen in younger people with some systemic illnesses such as chronic kidney disease, post mediastinal radiation and systemic inflammatory disorders like Takayasu arteritis, systemic lupus erythematosus and rheumatoid arthritis.[6–17]

Different authors, surgeons, forums or bodies have defined PA differently but the common denominator in all of them is that “aortic calcification” extends in such a manner that it interferes with aortic cannulation, aortic clamping, aortotomy and safe access to ascending aorta for AVR, necessitating modification of the surgical technique to avoid the complications. The valve consortium has defined it as severely atherosclerotic aorta characterized by heavy circumferential calcification or severe atheromatous plaques of the entire ascending aorta extending to the arch such that aortic cross clamping is not feasible.[18] CT scan is the imaging tool of choice to define PA [Fig. 1(A–C)].

A surgical classification given by Amorin et al, to guide treatment, consists of type 1 PA, when ascending aorta alone is involved and type II, if circumferential calcification involves descending thoracic aorta, with or without involving the arch. Type 1 is further classified as type 1 A, if it is impossible to cross-clamp the aortic root and as type 1B, if clamping is feasible but at a high surgical risk.[19]

PA is an incidental finding, mostly discovered during imaging studies carried out for cardiovascular and pulmonary diseases and the patients are asymptomatic with regards to PA. Prevalence of PA has been seen to be in a wide range of 2.7%–42.9% in people without cardiovascular disease. In patients undergoing electron beam CT for known or suspected CAD, the prevalence is reported to be 0.7%. Among patients with AS, PA is known to be present in approximately 7.5% subjects. In patients undergoing valve surgery or coronary artery bypass surgery, the prevalence of PA is reported to be 1.25–13%, whereas the same in those undergoing TAVR is in the range of 5%-33%.[20] However, PA can remain unnoticed many-a-times, only to be detected as a surgical finding after sternotomy.[21]

As mentioned above, PA may remain innocuous in itself, without producing any symptoms. However, a very stiff aorta can also lead to congestive heart failure, left ventricular (LV) failure, LV hypertrophy, elevated pulse pressure, coronary ischemia or sometimes, even sudden arrhythmic death.

Conventionally, before TAVR emerged as an alternative, surgery was the only option for those with severe AS, but in the presence of PA, the surgical AVR was either declined or the patient had no choice but to undergo an extensive and technically highly complex surgery generally associated with high rate of complications, particularly cerebrovascular embolic strokes. Two surgical alternatives to classical extensive surgery were attempted in these patients-surgery through axillary artery cannulation or, apico-aortic conduit or ascending aorta replacement. However, these clearly lacked efficacy and were associated with additional set of complications.[22–26] Sutureless AVR is another surgical advancement but its efficacy and safety in the presence of PA is still not proven.[27]

The advent of TAVR has allowed a promising therapeutic option for severe AS patients who also have PA. Currently in the era of TAVR, PA is an important independent factor for consideration during patient selection and is sometimes the sole indication for performing TAVR, irrespective of the risk category of aortic valve surgery. Fifteen percent of the inoperable patients in the randomized PARTNER study had PA. Data on TAVR in PA come from small series and case reports. A multicenter, observational, prospective study from three Spanish hospitals included 35 patients of severe AS with PA and their data showed no difference in terms of safety, feasibility and had similar rates of success and complications as compared to those seen in patients without PA. Axillary arterial access was chosen by some operators for patients who had unsuitable femoral access routes. Self-expanding Core Valve™ was used in the study.[28–31] Conventionally, TAVR for PA has been mostly done via transfemoral route, but some authors

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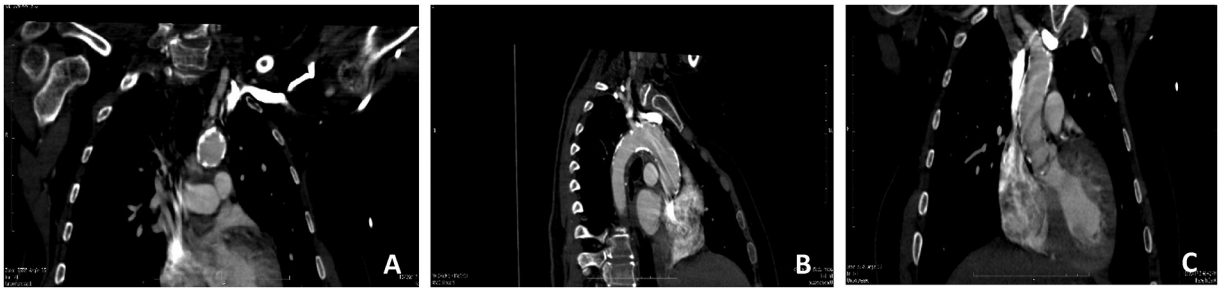

Fig. 1. (A,B,C): CT scan images of Porcelain Aorta showing partial and circumferential calcification of ascending aorta and the arch in a patient of severe aortic stenosis.

have shown good results with transapical approach as well, bypassing the whole aortic arch.[32]

With this background, we share our experience of TAVR in two cases of symptomatic severe calcific AS in whom PA was detected by the surgeons who subsequently declined to perform surgical AVR and the patients were referred for TAVR.

Interestingly, between the two patients, one was close to half of the other's age at the time of TAVR. The older patient, a 80-years old lady, had developed PA secondary to atherosclerotic intimal calcification, whereas the younger patient (age 38 years) had non-atherosclerotic medial calcification as the underlying pathology resulting from radiation therapy of the chest given at age five for Hodgkin's Lymphoma.

The first patient (the elderly lady) had previously undergone coronary artery bypass graft (CABG) surgery eight years back for CAD and her moderately stenosed aortic valve was not replaced by the surgeon because of PA and related risks of cross clamping during surgery. Post CABG, she was asymptomatic with her routine activities for few years. She then started having recurrence of angina. The grafts were patent but aortic stenosis had become severe. Surgical replacement of the valve was denied again because of the presence of PA, which was known since the past and further risks of redo open heart surgery and frailty at age of 80 years. Since TAVR had become available in India by that time, she underwent successful AVR by TAVR technique using a Core Valve™. The patient is doing well at six years follow-up after TAVR.

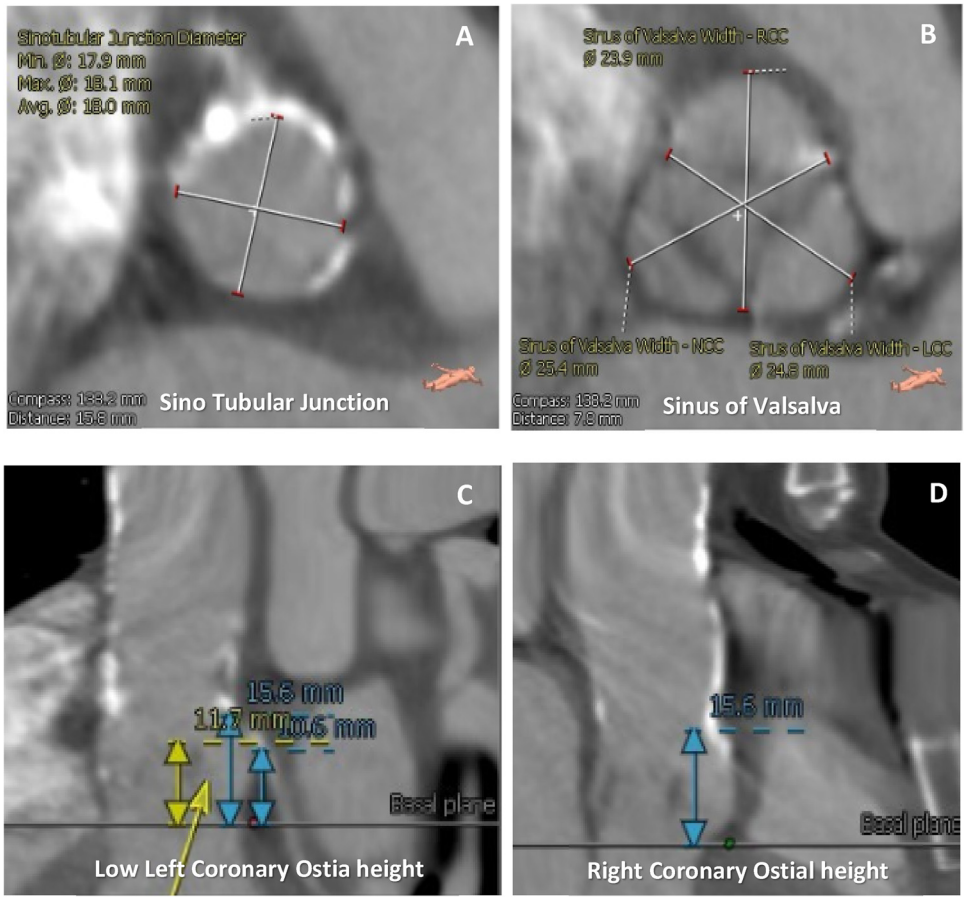

Fig. 2. (A,B,C,D): CT Images showing smaller sinus and low coronary ostia height.

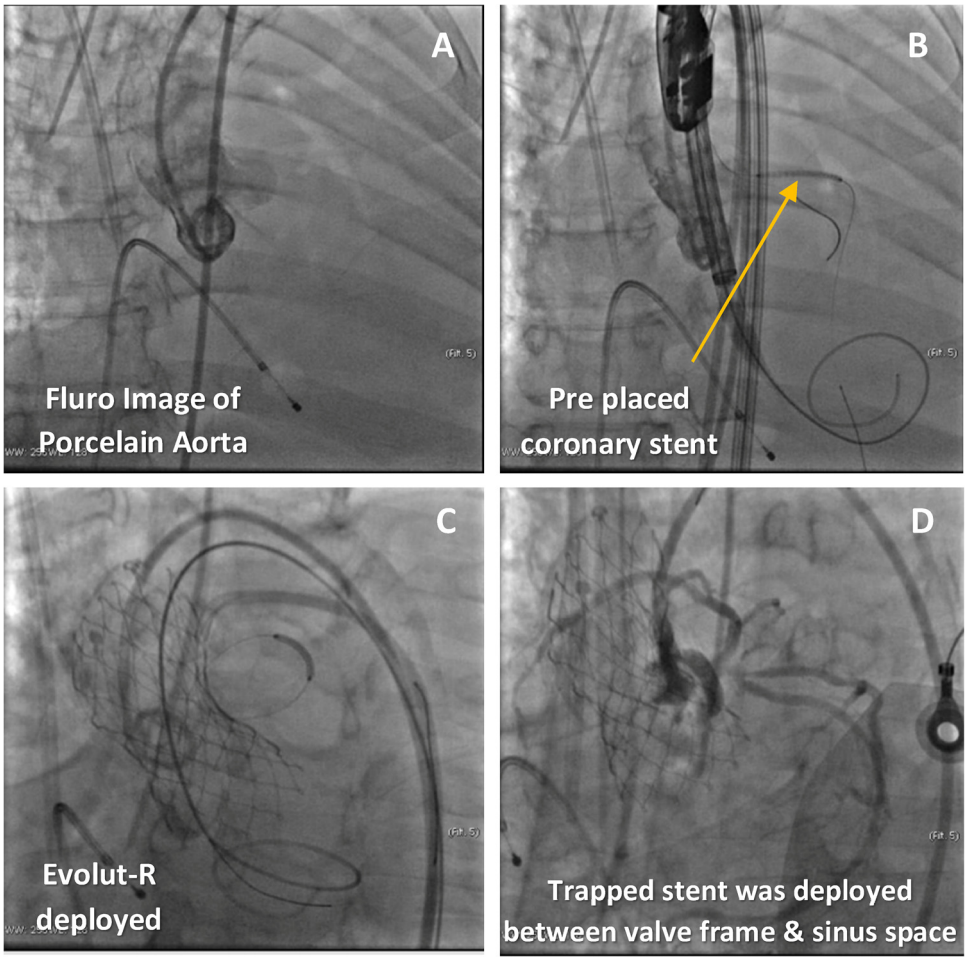

Fig. 3. (A,B,C,D): TAVR steps in Porcelain Aorta showing Coronary protection using pre placed Coronary Stent.

In contrast, the second patient is the youngest patient undergoing TAVR at our institution. As mentioned above, he was a 34-years old gentleman who had received radiotherapy for Hodgkin's lymphoma at five years of age. Irradiation of chest led to early degenerative changes and calcification of his aorta, aortic valve and coronary arteries. By 38-years of age, he had developed severe, symptomatic calcific AS. Breathlessness was the main symptom and he was in New York Heart Association functional class III at the time of presentation. The cardiac surgeons declined to perform a surgical AVR on him due to PA observed on multi-slice CT scan and therefore, he was advised TAVR. The pre-TAVR work up revealed heavily calcified tricuspid aortic valve, calcified ascending aorta and sinotubular junction, and small sinuses of Valsalva. The height of left coronary arteries ostia was 11.7 mm and the right was 13.4 mm [Fig. 2(A–D)]. The sinus of Valsalva diameters were $24.8 \times 23.9 \times 25.4$ mm and the sinotubular junction diameter was 17.9×18.1 mm [Fig. 2(A–D)]. In view of lower left coronary ostia height and small sinuses, there was concern about the pinching of left main ostia. Coronary protection strategy was planned. A coronary stent was pre-placed in the left anterior descending artery over an All-star coronary wire [Fig. 3(A–D)]. A 23-mm Evolut R^{TM} aortic valve was then deployed successfully and the coronary stent was pulled out once no coronary impingement was confirmed after deployment of the aortic prosthesis on an aortogram done post implantation. Unfortunately, the stent got trapped between the sinus of Valsalva and the prosthesis frame. Different maneuvers were tried to pull out the trapped stent but to no result. A new strategy was then applied wherein a chimney between the frame of device, sinus and the left main coronary artery was created by deploying the stent in the potential space much away from the ostium of left main [Fig. 3(A–D)]. Coronary flow to the left system was not compromised and TAVR was completed successfully. The patient was doing well at one-month follow-up after the procedure and the follow-up CT scan revealed well deployed Evolut-R valve, patent stent (chimney like) and unimpaired left main coronary flow [Fig. 4(A–C)].

Thus, to conclude, given its technical feasibility, TAVR is likely to become the preferred strategy for treating patients with severe AS and concomitant PA. PA is an established impediment to surgical AVR because of the technical inoperability and is associated with increased risk of stroke. The other surgical alternatives for severe AS in presence of PA are technically very complex and challenging. In comparison, TAVR is a much safer and easier option for treating such patients. Advancements in TAVR technology have made it possible that whether it is a hell or high water arising from the burden and distribution of calcium in PA, TAVR can navigate and reach the shore in most patients with much lesser risk of complications!

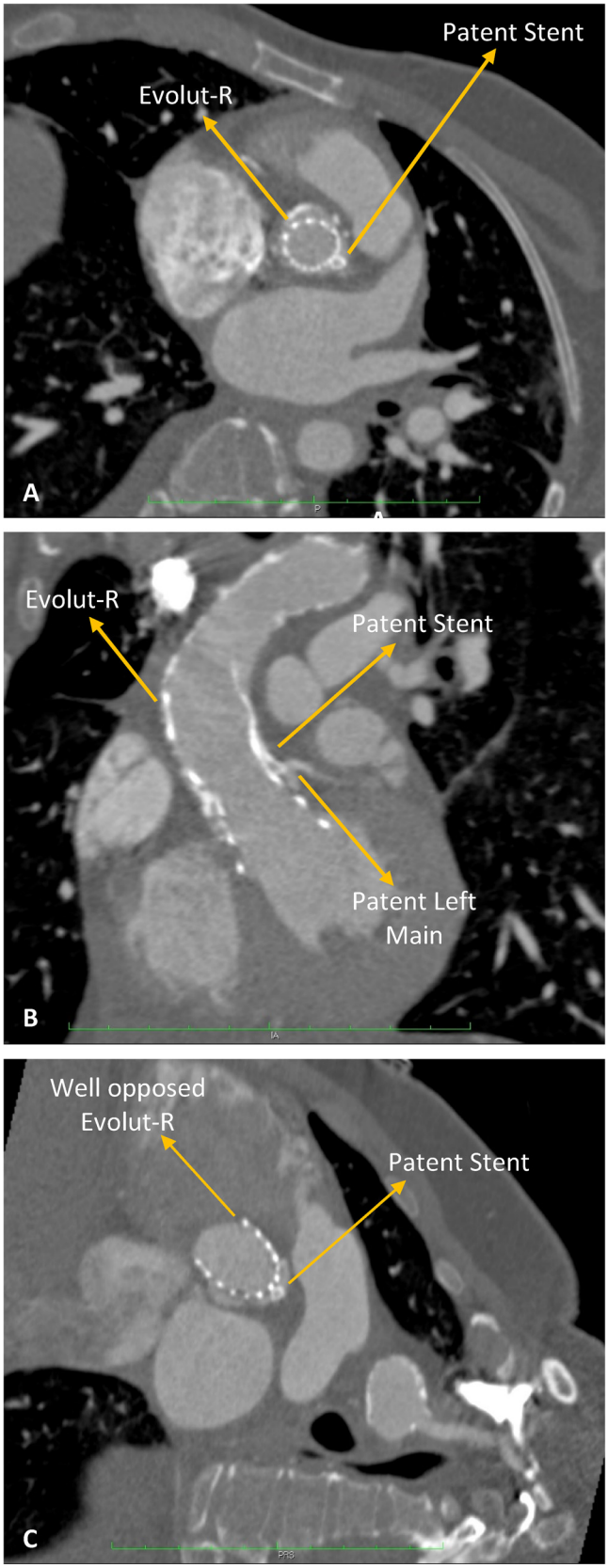

Fig. 4. (A,B,C): CT Aortogram at 1-month follow-up shows well deployed Evolut-R & patent stent.

Conflict of interest

None.

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