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CASE REPORT

INTERMEDIATE

CLINICAL CASE

# Unusual Manifestations of Coral Reef Aorta Complicated Severe Valvular Heart Disease in a Dialysis Patient

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## ABSTRACT

We report the unusual case of a 68-year-old man with coral reef aorta complicated with severe calcified valvular heart disease who has been undergoing dialysis for 21 years. This report highlights the etiology and the unusual manifestations of coral reef aorta in a long-term dialysis patient. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2021;3:1705–1710) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

## HISTORY OF PRESENTATION

A 68-year-old man with end-stage renal disease (ESRD) requiring dialysis was admitted for intradialytic hypotension (IDH) and a month history of progressive rest pain on the right lower extremity. Five months earlier, the patient had been diagnosed with IDH and treated with amezinium methylsulfate at another facility. On presentation, physical examination on a nondialysis day revealed a painful nonhealing ulcer on the right great toe with absent bilateral femoral pulses. Blood pressure was 93/58 mm Hg with a regular pulse rate of 67 beats/min, and the respiratory rate and oxygen saturation were within normal ranges. Cardiovascular examination revealed a grade 3 systolic ejection murmur and a single second heart sound, and the patient's tibial arteries had only weak Doppler signals.

### LEARNING OBJECTIVES

- To recognize severe obstructive calcification of the aorta as a complication of ESRD-related vascular changes.
- To demonstrate the adverse effects of VHD and LVH, which predispose to IDH, in a long-term dialysis patient.

## PAST MEDICAL HISTORY

The patient had a history of chronic glomerulonephritis complicated by ESRD requiring dialysis for the previous 21 years. The patient had been prescribed calcium carbonate for hyperphosphatemia. Nine years before the presentation, the patient's cardiovascular history included endovascular therapy and femorofemoral bypass for severe occlusive disease of both common iliac arteries. Since then, the patient had ceased smoking, and his cholesterol level had been managed with statins.

## DIFFERENTIAL DIAGNOSIS

Differential diagnoses for critical limb ischemia (CLI) included progressive peripheral artery disease or a complication of IDH.

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**ABBREVIATIONS AND ACRONYMS**

**CLI** = critical limb ischemia

**CRA** = coral reef aorta

**CTA** = computed tomography angiography

**ESRD** = end-stage renal disease

**IDH** = intradialytic hypotension

**LVH** = left ventricular hypertrophy

**VHD** = valvular heart disease

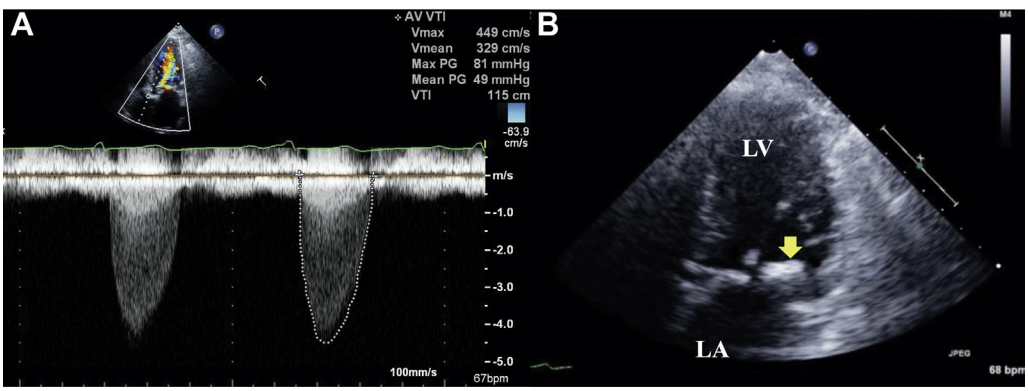
## INVESTIGATIONS

The ankle-brachial index was unmeasurable, and the skin perfusion pressures of the foot were 14 mm Hg. Serum examination showed a calcium level of 9.2 mg/dL (normal range 8.6-10.2 mg/dL), a phosphorus level of 4.4 mg/dL (normal range 2.5-4.5 mg/dL), and a low-density lipoprotein cholesterol level of 72 mg/dL (normal range 65-163 mg/dL). Transthoracic echocardiography demonstrated concentric left ventricular hypertrophy (LVH), severe aortic stenosis, and moderate mitral stenosis. The interventricular septal diastolic thickness was 1.2 cm, and the left ventricular posterior wall thickness was 1.3 cm. The patient had a preserved ejection fraction of 72% and a mean gradient of 49 mm Hg across the aortic valve with an aortic valve area of 0.76 $cm^2$. Moderate mitral stenosis with a mean pressure gradient of 7.0 mm Hg and a mitral valve area of 1.48 $cm^2$ was also identified (**Figure 1**). Cardiac computed tomography revealed C-shaped mitral annular calcification connecting to the left ventricular outflow tract calcification and severe calcified aortic valve (**Figure 2,** Video 1). Computed tomography angiography (CTA) showed an extensively calcified atheromatous aorta distal to the celiac artery. The calcifications protruded and occupied the lumen of the aorta (**Figure 3**, Video 2). The features of CTA consisted of a coral reef aorta (CRA) (1,2). The prior femorofemoral bypass was patent, and the infrainguinal peripheral arteries had no significant stenosis. The coronary angiography revealed no significant stenoses regardless of the high Agatston calcium score of 1,862.

## MANAGEMENT

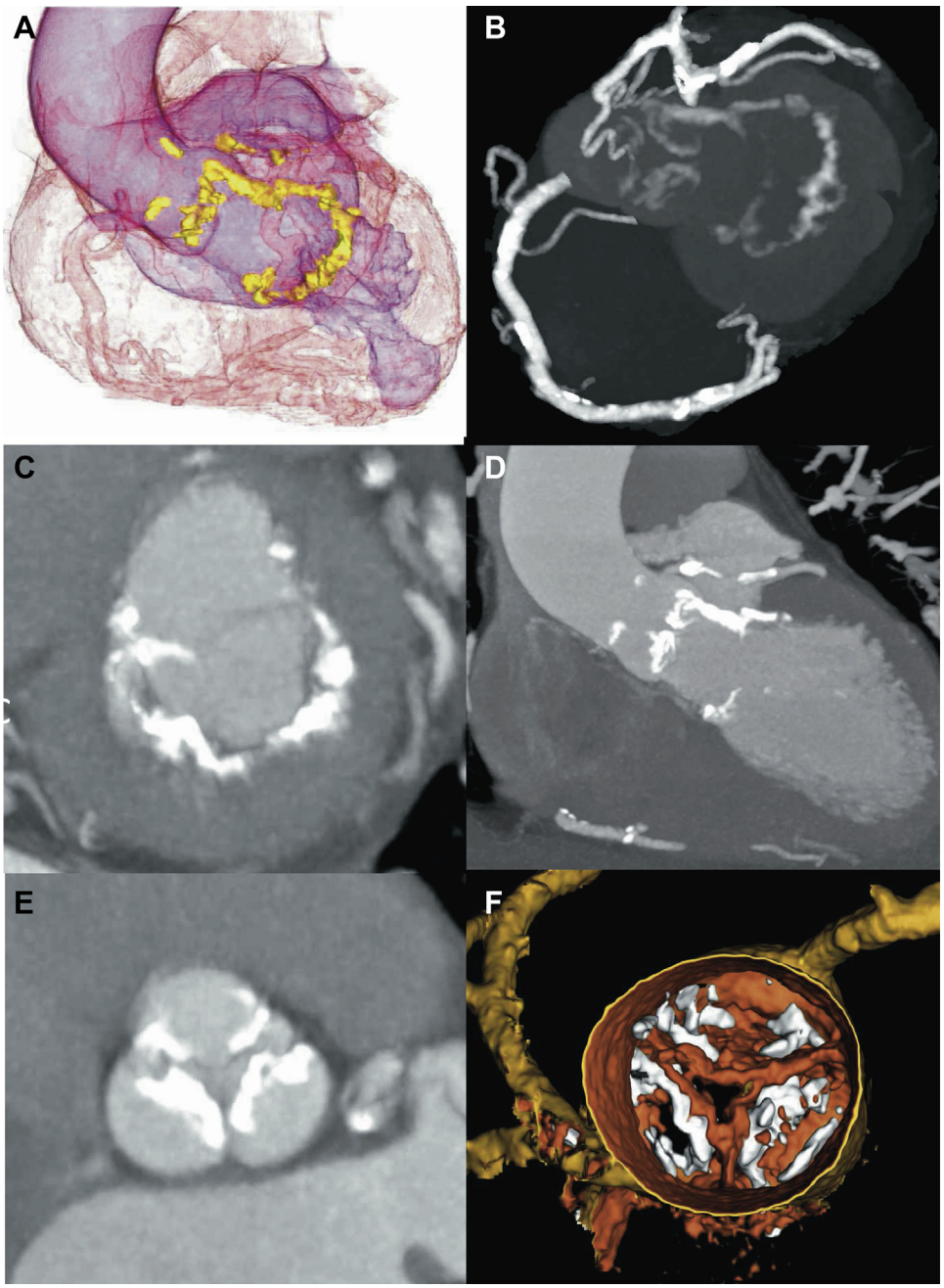
Following discussion among the members of the multidisciplinary cardiovascular team, the decision for concomitant surgical valve replacement and surgical revascularization was made considering severe valvular heart disease (VHD) and CLI with a moderate risk of amputation according to the Wound, Ischemia, and Foot Infection classification system in compliance with the current guidelines (3,4). Transcatheter aortic valve replacement was contraindicated because of the surgically treatable mitral stenosis (3). Intraoperatively, standard median sternotomy was performed, and simultaneously, the middle part of the prosthetic graft of the femorofemoral bypass was exposed. A 10-mm prosthetic graft served as a subcutaneous conduit from the pericardial space to the exposed graft. One end of the prosthetic graft was anastomosed to the exposed graft, and the other end of the prosthetic graft was connected to the cardiopulmonary bypass circuit for the lower body perfusion. After cannulating an ascending aorta for upper body perfusion and bicaval venous drainage, mitral valve replacement preserving a posterior leaflet was completed with an Epic bioprosthesis (St. Jude Medical) followed by aortic valve replacement of an INSPIRIS RESILIA valve (Edwards Lifesciences) (**Figure 4**). Then, the extra-anatomic ascending aorta to bifemoral bypass was completed by anastomosing the proximal end of the prosthetic graft. The

**FIGURE 1 Transthoracic Echocardiography Images**




**(A)** Transthoracic echocardiographic images demonstrating severe aortic stenosis. **(B)** Four-chamber view of the mitral valve demonstrating severe mitral annular calcification, which engulfed the posterior leaflet **(yellow arrow)**. LA = left atrium; LV = left ventricle.

**FIGURE 2** **Cardiac Computed Tomography Images**




**(A)** Three-dimensional image of the heart **(yellow)** and **(B)** the maximum intensity projection demonstrating C-shaped mitral annular calcification connecting to the left ventricular outflow tract calcification and calcified aortic valve. **(C)** Short-axis view of the mitral valve demonstrating severe mitral annular calcification. **(D)** Coronal view of the left ventricular outflow tract demonstrating severe calcification. **(E)** Short-axis view and **(F)** 3-dimensional image of the aortic valve demonstrating severe calcification.

postoperative course was uneventful. Hemodynamic status during maintenance dialysis became stable, and the ischemic ulcer on the right great toe was healed without amputation. The skin perfusion pressures of the foot recovered to 84 mm Hg. CTA demonstrated that the bloodstream of the right lower extremity depended on the patency of the bypass (Figure 5).

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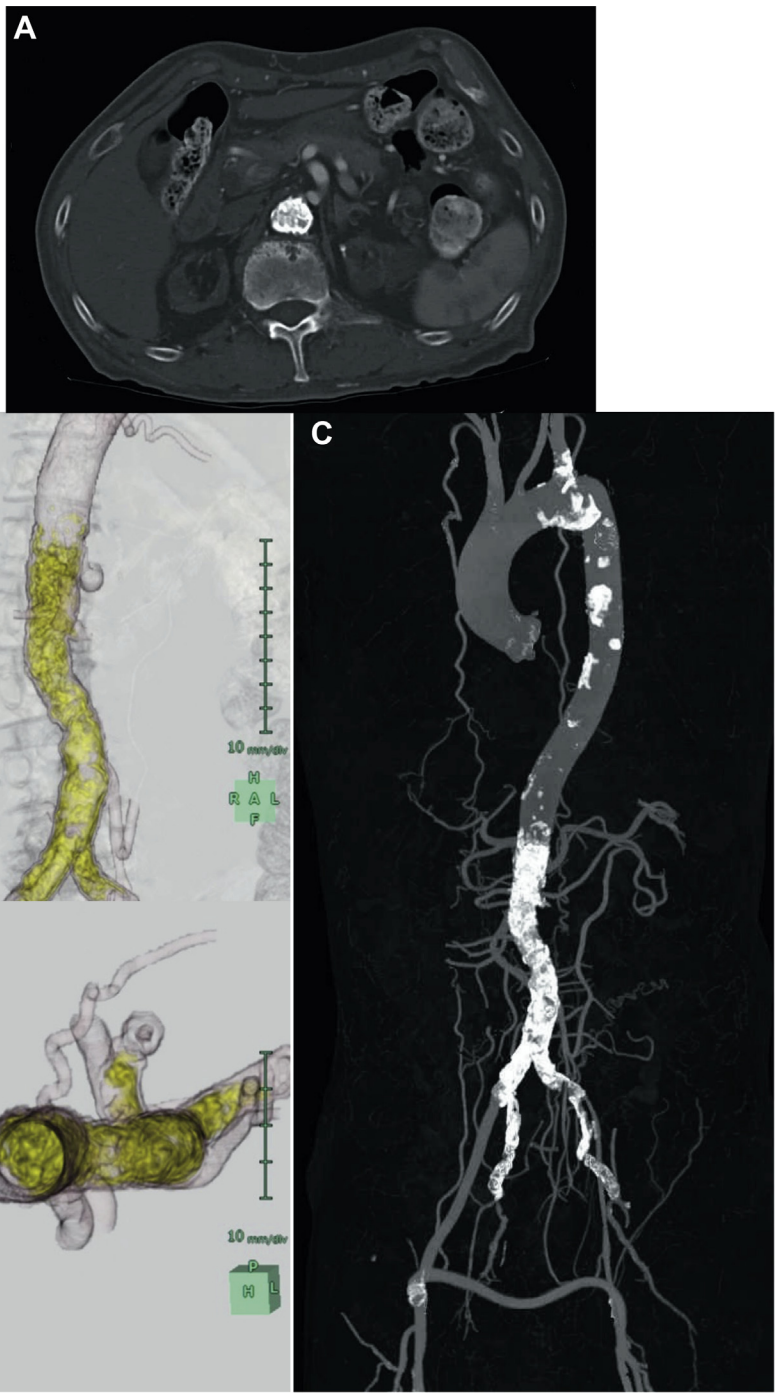
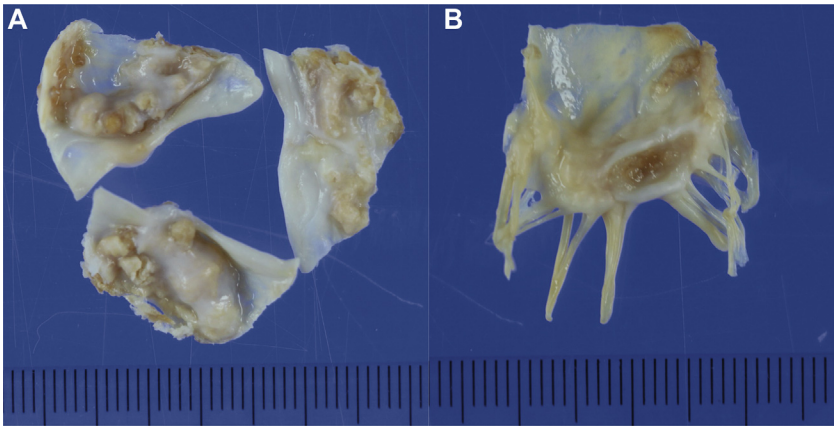
**FIGURE 3 Computed Tomographic Angiogram Images**




**(A)** Computed tomographic angiogram and **(B)** 3-dimensional images demonstrating an extreme calcified atheromatous plaque that protruded into the aortic lumen just below the celiac artery. **(C)** Maximum intensity projection demonstrating occlusive calcified thoracoabdominal aorta from distal to the celiac artery to both common iliac arteries.

**FIGURE 4 Macroscopic Images of the Specimens**




**(A)** Aortic valve specimen and **(B)** anterior mitral leaflet specimen view showing heavily calcified change.

## FOLLOW-UP

After 2 years of surgery, no cardiovascular event was reported.

## DISCUSSION

CRA is a rare and unique entity of extensive calcification causing significant stenosis of the aorta and has a high operative mortality rate of 8.7% to 11.6% (1). The cardiovascular changes in ESRD patients leading to high mortality rates include LVH and accelerated atherosclerosis and arteriosclerosis that are characterized by diffuse calcification (5). Calcification of cardiac valves, including mitral annular calcification, is also well known to occur in patients with ESRD. To our best knowledge, this case is the only presentation of CRA complicated with severe VHD in a long-term dialysis patient (1,2). The etiology and pathogenesis of CRA remain unclear (1,2). Calcification with increased arterial wall thickness can be found in a large elastic artery, as well as a specific ESRD-related vascular calcification in small to medium arteries. The mechanisms of vascular calcification include passive precipitation of calcium and phosphate, which affects osteogenic transformation and hydroxyapatite formation, and deficiency of calcification inhibitors (6). Several factors may lead to severe calcification of the cardiac valves and aorta. Calcium and phosphate metabolism related to long-term dialysis, including hyperphosphatemia treated with calcium carbonate, may have contributed to the development of CRA and severe VHD in this case.

**FIGURE 5 Postoperative 3-Dimensional Computed Tomographic Angiogram Image**

Postoperative 3-dimensional computed tomographic angiogram confirming the patency of the extra-anatomical bypass from the ascending aorta to the femoral arteries.

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The manifestations of CLI following IDH are also distinct. Most patients with CRA have been reported to have refractory hypertension, intermittent claudication, and symptoms associated with visceral ischemia (1,2). Aortic occlusive disease without the infrainguinal disease rarely causes CLI. IDH, to which ESRD-related LVH and VHD predispose (7), can contribute to the manifestation of CLI.

## CONCLUSIONS

CRA is a potential complication in long-term dialysis patients. We must be aware of the adverse effects of ESRD-related cardiovascular complications, such as LVH and VHD, which predispose to IDH. The timely surgical treatment with a multidisciplinary approach is imperative for treating these complications.

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**KEY WORDS** coral reef aorta, dialysis, end-stage renal failure, intradialytic hypotension, valvular heart disease

**APPENDIX** For supplemental videos, please see the online version of this paper.