

# A BLACK PERCUTANEOUS CORONARY INTERVENTION AND TRANSCATHETER AORTIC VALVE IMPLANTATION

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## HISTORY AND PHYSICAL

A 78 year-old female was referred for inferior ST-elevation-myocardial-infarction and syncope (Panel-A). Physical examination revealed a 3/6 diffuse systolic ejection murmur, unusual dark-spots of the eyes and auricular and nails grayish-bluish discoloration (Panel B-F). Echocardiography revealed severe aortic stenosis, akinesia of the inferior wall with a left ventricular ejection fraction (LVEF) of 38% (Panel F-G)

## IMAGING

Urgent coronary angiography showed occlusion of the right coronary artery (RCA), (Panel H), critical stenosis of the left main and the left anterior descending artery (LAD) (Panel-I). Primary-percutaneous coronary intervention (PCI) on RCA was performed with satisfactory result (Panel-k). Further, she suffered of a debilitating diffuse arthropathy with a previous history of a bilateral hip prostheses (Panels M-P). A diagnosis of alkaptonuria was done. Due to high surgical-risk, a CT evaluation pre-Transcatheter Aortic Valve Implantation (TAVI), (Figure 2 Panel-A) was performed. An IVUS-guided unprotected left main-LAD PCI with drug-eluting-stent implantation was successfully performed (Figure 2 Panel-B-D).

## INDICATION FOR INTERVENTION

However, patient became hemodynamically unstable and symptomatic for presyncope episodes and acute heart failure.

## INTERVENTION

An emergency balloon aortic valvuloplasty (BAV) was performed with complete symptoms relief and LVEF recovery (Figure 2- Panel E). Patient was discharged in good condition with indication to perform TAVI and a strict follow-up. Notably, due to symptom control, after 20-months, a 29mm Corevalve prosthesis (Figure 2- Panel F) was implanted with satisfactory result and residual mild paravalvular aortic regurgitation.

## LEARNING POINTS

Ochronosis is a heritable-disorder of tyrosine metabolism, with various systemic abnormalities related to pigment deposition and degeneration of collagen and other tissues. Cardiovascular manifestations of alkaptonuria often include aortic stenosis and rarely coronary artery disease.

An “invasive-minimalist” therapy can be the best treatment option in frail patients with ochronosis due to the presence of hostile chest due to extensive calcifications and porcelain aorta.

To the best of our knowledge, this case represents the first description of a Percutaneous Coronary Intervention (PCI), aortic balloon aortic valvuloplasty (BAV) and Transcatheter Aortic Valve Implantation (TAVI) in a patient affected by ochronosis, with high rates of primary success.

We believe that the illustration of our case might contribute to a better management of these frail patients at elevated cardiovascular-surgery risk, in which PCI, balloon aortic valvuloplasty and TAVI may represent viable therapeutic options.

