

# Transradial Coronary Angioplasty in an Achondroplastic Patient with Chronic Total Occlusion (CTO): First Case Report

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## ABSTRACT

Coronary interventions in patients with achondroplasia have been very rarely reported. The optimal approach to such patients in complex cases is not known. They pose special challenge to coronary revascularisation due to short stature, angle of their elbows, kyphoscoliosis, frequent obesity, and lack of specific equipments and experience. We present an interesting case of 67 years old man with achondroplasia who presented with significant angina symptoms and past history of IWMI. Transradial angiography showed chronic total occlusion (CTO) of right coronary artery. Coronary angioplasty was done via right radial approach using available hardware with good results. To our knowledge, this is the first case of radial angioplasty in an achondroplastic patient with CTO. The procedural difficulties and techniques have been discussed along with brief review of literature.

**Key words:** Chronic stable angina, Dwarf, Kyphoscoliosis, Percutaneous coronary intervention, Short stature.

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## INTRODUCTION

Achondroplasia is an autosomal dominant condition caused by a mutation in fibroblast growth factor receptor 3 (FGFR3).<sup>1</sup> It is most common dwarfing condition having a prevalence of 1/25,000 live births. Mortality is 2.27 times more than that of the general population, cardiovascular problems being the most frequent cause of death. The average life expectancy of this population is decreased by 10 years.<sup>2</sup> Obesity begins in early childhood and is prevalent at all ages.<sup>3</sup> Characteristic features include an average-size trunk, short arms and legs, with particularly short upper arms and thighs and limited range of motion at the elbows.<sup>4</sup>

Despite higher prevalence of heart disease in this population, there is scarcity of data on coronary interventions in such patients. The optimal approach to such patients in complex cases is not known. We hereby report an interesting case of an achondroplastic patient who presented with significant angina symptoms and CTO (Chronic Total Occlusion) of RCA (Right Coronary Artery). He underwent successful radial angioplasty with good results and no complications.

## CASE REPORT

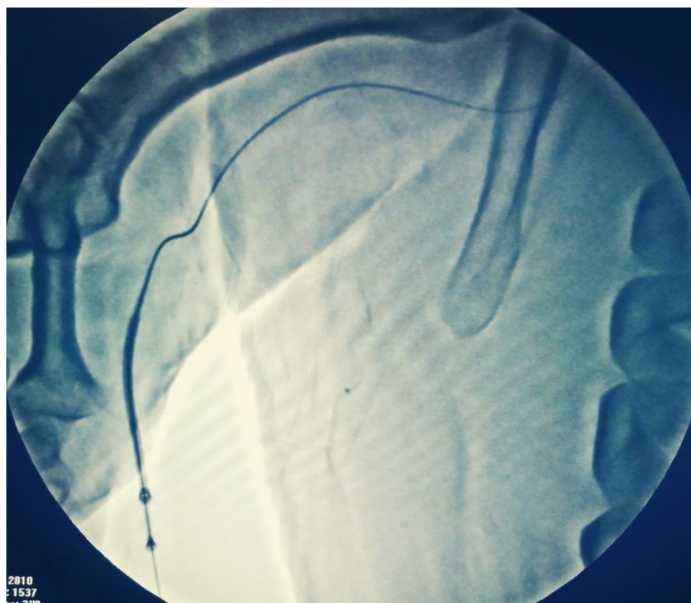
A 67-year old Indian male with achondroplasia presented with chronic stable angina CCS (Canadian Cardiovascular Society) class III since previous 6 months. He had suffered inferior wall myocardial infarction (IWMI) two years back. He was a chronic heavy smoker and also had dyslipidemia and obesity as coronary risk factors. He had very short limbs and severe kyphoscoliosis (Figure 1). The ECG (Electrocardiogram) showed Q wave in inferior leads with T wave inversion. Echocardiography revealed RWMA (Regional Wall Motion Abnormality) in RCA (Right Coronary Artery) territory with LVEF (Left Ventricular Ejection Fraction) of 45%.

Coronary angiography was planned in view of significant angina symptoms despite maximum medical management. Right radial artery approach was chosen in view of femoral artery access issues, to avoid local bleeding complications and early mobilisation. Initially radial artery sheath length was compared to total upper limb length to ensure that radio-brachial system was long enough to accommodate the radial sheath. A 6F radial artery sheath was then introduced only half way and

sheath angiogram was taken to rule out any arterial anomaly in radio-brachial system. Thereafter a terumo wire was passed all the way to heart to have an overview of tortuosity in the arterial system and patency. Once found normal the sheath was pushed fully inside beyond elbow joint just falling short of shoulder joint (Figure 2). Coronary angiography was done using 5F Tiger catheter with the major portion of the catheter lying outside body (Figure 3). Tracking the diagnostic catheter was very difficult due to severe radio-brachial tortuosity. Cannulation of coronary arteries was also difficult *and asking patient to hold breath in inspiration was helpful in cannulation. Angiographic views had to be modified to attain classical views. Angiogram showed total occlusion of the*



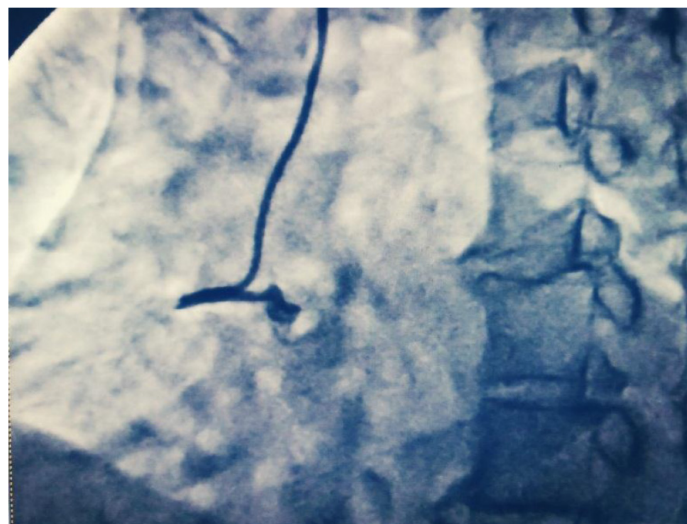
**Figure 1:** The patient of achondroplasia.



**Figure 2:** The radial sheath in situ, seen well beyond elbow joint just falling short of shoulder joint. Note the tortuosity in the arterial system.



**Figure 3:** The procedure was performed with most of the catheter length lying outside body.



**Figure 4:** Coronary angiogram: Right Coronary Artery (RCA) proximal chronic total occlusion (CTO).



**Figure 5:** Post coronary angioplasty: stenting to RCA with Xience Expedition 3x38mm stent, TIMI 3 flow.

proximal RCA with normal left coronary system in a right dominant system (Figure 4). PCI (Percutaneous Coronary Intervention) was planned after taking informed consent.

The 5F Tiger diagnostic catheter was exchanged over 0.032" Teflon wire for 6F, 3.5 curve Judkins Right (JR) guiding catheter. JR guiding catheter yielded good support after hooking RCA due to highly tortuous arterial anatomy. Initial crossing of lesion was unsuccessful with 0.014" BMW (balance middle weight) and Fielder XT guidewire. Lesion was finally crossed with 0.014" Cross IT 100XT wire with Fine Cross microcatheter support. Serial pre-dilatations were done using 1.5x10mm, 2x10mm and 2.5x10mm semi-compliant balloons. The RCA lesion was stented with Xience Expedition 3x38 mm (drug eluting stent) at 14 atm. Post-dilatation was done sequentially using 3.5 x 12 mm NC Sprinter (Non-Compliant) balloon from 14 to 18 atm. Distal TIMI 3 flow (Throm-

bolysis In Myocardial Infarction) was achieved at the end of the procedure without any complications (Figure 5).

The course in the hospital was uneventful and patient was discharged in a stable condition on second day of hospitalization. At discharge, the patient was put on aspirin, clopidogrel, statin, beta-blocker and ACE inhibitor. He is on regular follow up without any symptom since last 6 months.

## DISCUSSION

Achondroplasia is the most common cause of dwarfism in patients with short limbs.<sup>1</sup> The overall heart disease mortality rate is over two-fold greater than that of the general population.<sup>2</sup> Despite increased prevalence of heart disease, the optimal approach of revascularisation in such patients is not known due to scarcity of literature. CABG (Coronary artery bypass Graft) is many times limited by non-availability of normal

saphenous vein grafts. However, there are a few reports of coronary artery bypass surgery in these patients, including the use of both a saphenous vein and the left internal mammary artery,<sup>5</sup> report of successful CABG after a phlebography of the limbs revealed adequate saphenous veins,<sup>6</sup> and CABG using one saphenous vein graft and bilateral internal mammary artery grafts.<sup>7</sup>

There are also few reports of PCI including radial PCI to RCA as rescue PCI post thrombolysis,<sup>8</sup> Sheathless transradial angioplasty to LAD (Left Anterior Descending artery) in STEMI (ST Elevation Myocardial Infarction),<sup>9</sup> and multivessel femoral PCI.<sup>10</sup> Our case of transradial angioplasty differs from earlier reports because this involves complex PCI. Moreover the technical details and difficulties have been described in greater detail.

The relative indications and risks of radial versus femoral approach are not known because of lack of literature. However, wherever possible radial artery approach may be preferred in view of femoral artery access issues, to avoid local bleeding complications and early mobilisation. The best approach and complication rates in achondroplasia population would require further investigations. Performing coronary interventions in such patients is markedly challenging, especially through the transradial route, because of their short limbs, angle of their elbows and kyphoscoliosis.

To best of our knowledge, this is first case of complex PCI in an achondroplastic patient using transradial route. We have hereby demonstrated that available hardware can be used to perform even complex angioplasty in such patients with transradial approach. Severe technical difficulties were encountered while negotiating the catheter and trying to cannulate the coronary arteries. Crossing the lesion was also challenging, but was successful with wire escalation. Further studies need to be done to identify the best approach and improve the technical aspects of the procedure in these patients.

## CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

## ABBREVIATIONS

**IWMI:** Inferior Wall Myocardial Infarction; **CTO:** Chronic Total Occlusion; **RCA:** Right Coronary Artery; **CCS:** Canadian Cardiovascular Society; **RWMA:** Regional Wall Motion Abnormality; **LVEF:** Left Ventricular Ejection Fraction; **PCI:** Percutaneous Coronary Intervention; **CABG:** Coronary Artery Bypass Graft; **STEMI:** ST Elevation Myocardial Infarction.

## SUMMARY

Despite higher prevalence of heart disease, there is scarcity of data regarding coronary intervention in achondroplastic patients. The optimal approach in complex coronary lesions is not known in these patients. We have reported successful trans-radial coronary angioplasty in a patient with chronic total occlusion (CTO) using available hardware. Radial approach therefore can be considered in the patients with achondroplasia, however further studies are required to define the best approach.

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