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Case Report

Survival Against the Odds: An Exceedingly Uncommon Case of Type A3 Truncus Arteriosus in an Adult Male Thriving without Surgical Intervention

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ABSTRACT

Persistent truncus arteriosus is an uncommon complex congenital heart disease. Without surgical intervention, the prognosis is not good. Very few untreated cases of truncus arteriosus survive to maturity. We report an exceptional case of uncorrected truncus arteriosus surviving into adulthood of Type A3 variant -Van Praagh classification as a 26-year-old male who had well tolerated dyspnea since birth. Our patient showed a single(right) pulmonary artery arising from the common arterial trunk supplying the right lung and absent main and left pulmonary artery with collateral supply to the left lung consistent with the truncus arteriosus Type A3-Van Praagh classification

Keywords: Persistent truncus arteriosus; Congenital heart disease; Surgical intervention; Van Praagh; Arterial trunk

Introduction

Truncus arteriosus usually presents early after birth and accounts for 1% of congenital heart disease[1]. It is typified by the truncus arteriosus failing to split during fetal development into the pulmonary and aortic arteries, resulting in a single artery with different properties. Among the four subtypes of truncus arteriosus, type IV has a relatively longer survival rate due to low pulmonary blood flow, while types I, II and III have significantly worse outcomes, with 80% of infants dying before reaching infancy. This is because of the increased incidence of heart failure due to higher pulmonary blood flow, which causes mortality[2].

Since the prognosis for types 1 - 3 is so bad, very little is known about their natural, unrepaired course. However, there have been very few cases of survivorship beyond infancy without intervention[3]. Echocardiography might not always be helpful in assessing this kind of CHD. Recognition of this CHD is facilitated by its presence on computed tomography (CT). We talked about the CT imaging features of case of type A3 truncus arteriosus in our case report.

Case Presentation

A 25-year-old male presented to cardiology OPD at Jawaharlal Nehru Medical College, a tertiary care hospital, with complaints of well-tolerated breathlessness since childhood. He weighed 40

kilograms and stood 156 cm tall. Evidence of clubbing with no cyanosis. His blood pressure was 128/64 mmHg and his heart rate was 88/min. The SpO2 measurement using pulse oximetry was 98% and pansystolic murmur on auscultation was recorded. Echocardiography revealed a single common arterial trunk, large mal-aligned VSD with mild AR and a diagnosis of hemi-truncus was made[4].

A multi-phasic cardiac CT demonstrated cardiomegaly with a single common arterial trunk or truncus arteriosus **(Figure 1) and** an only pulmonary artery emerging from the postero-lateral aspect of common arterial trunk above the valvular level was noted to have tortuous course and ultimately supplying the right hilum i.e. right pulmonary artery **(Figures 2a& 2b)**. Main pulmonary artery and pulmonary artery for left hilum were absent. Two MAPCAs (major aortopulmonary collateral arteries) supplied the right lung **(Figures 3,4)**, while the left lung was supplied by few minor collaterals **(Figures 5,6)**. An associated large membranous ventricular septal defect was noted **(Figures 7,8)**.

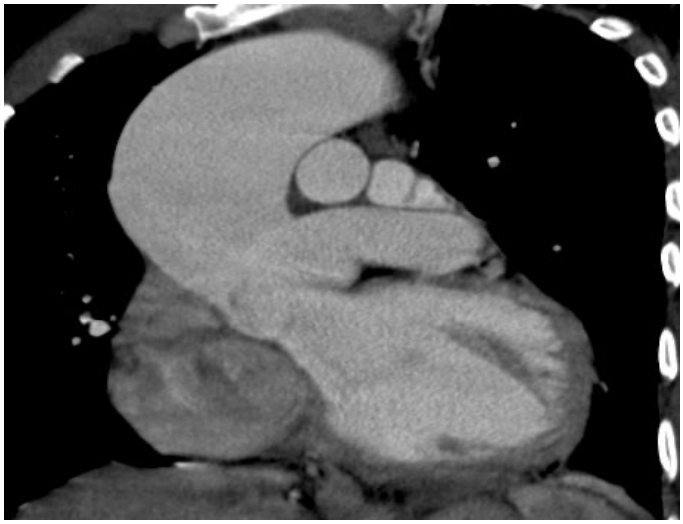

Figure 1: Coronal CT scan shows a common arterial trunk arising from left ventricle with over-riding of aorta and VSD.

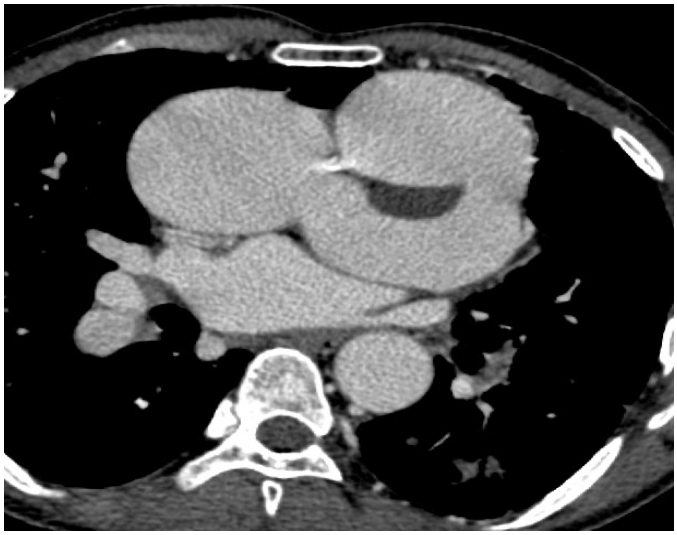

Figure 2a: Axial CT: A pulmonary artery arising from the posterolateral aspect of common arterial trunk (Truncus) from its posterolateral aspect and continuing as right pulmonary artery supplying right hilum.

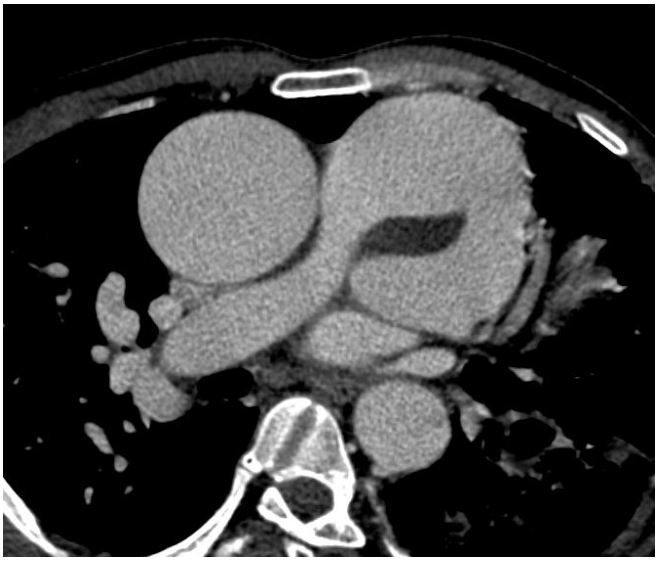

Figure 2b: Axial CT: A pulmonary artery arising from the posterolateral aspect of common arterial trunk (Truncus) from its posterolateral aspect and continuing as right pulmonary artery supplying right hilum.

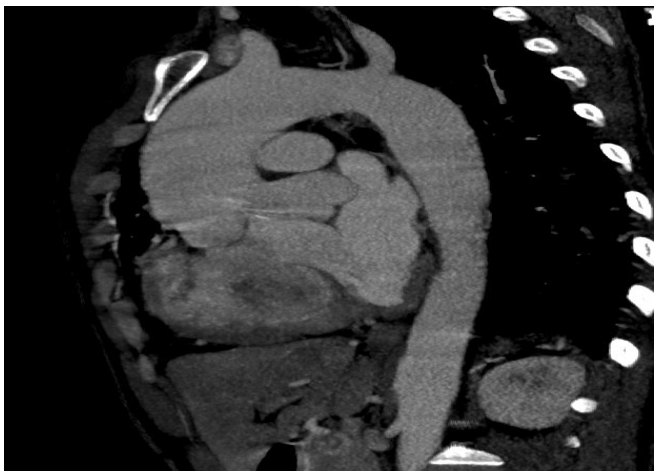

Figure 3: Axial CT: A pulmonary artery arising from the posterolateral aspect of common arterial trunk (Truncus) from its posterolateral aspect and continuing as right pulmonary artery supplying right hilum.

Figure 4: Axial CT: A pulmonary artery arising from the posterolateral aspect of common arterial trunk (Truncus) from its posterolateral aspect and continuing as right pulmonary artery supplying right hilum.

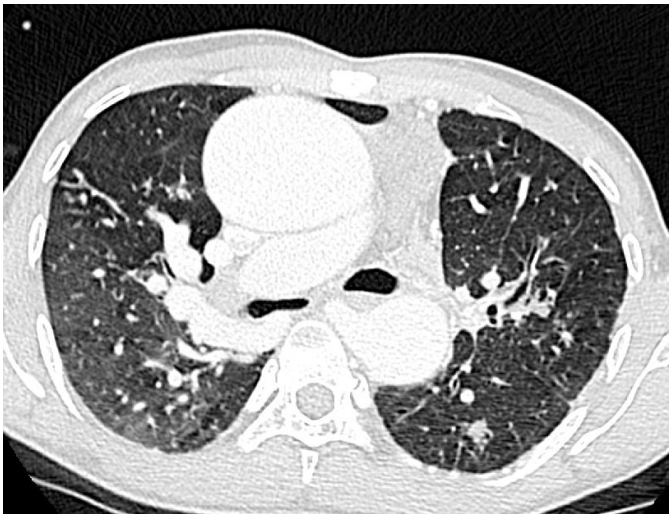

Figure 5: Axial CT: A pulmonary artery arising from the posterolateral aspect of common arterial trunk (Truncus) from its posterolateral aspect and continuing as right pulmonary artery supplying right hilum.

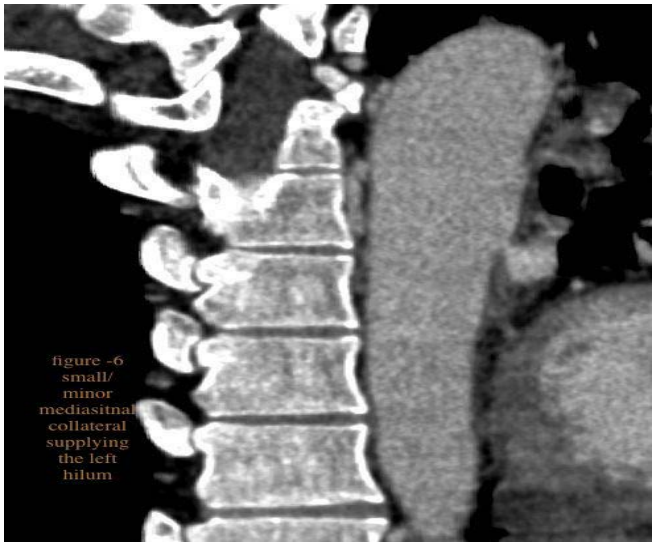

Figure 6: Axial CT: A pulmonary artery arising from the posterolateral aspect of common arterial trunk (Truncus) from its posterolateral aspect and continuing as right pulmonary artery supplying right hilum.

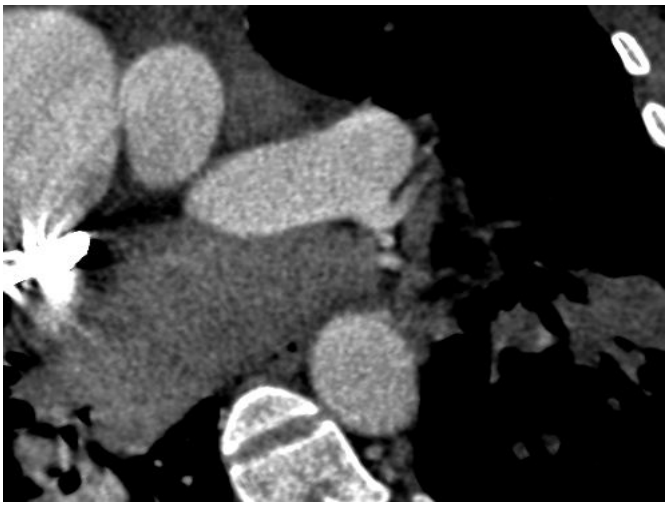

Figure 7: Axial CT: A pulmonary artery arising from the posterolateral aspect of common arterial trunk (Truncus) from its posterolateral aspect and continuing as right pulmonary artery supplying right hilum.

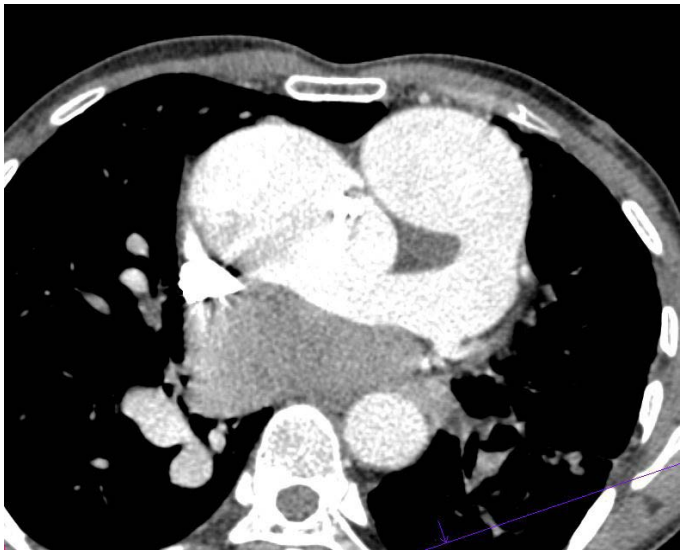

Figure 8: Axial CT: A pulmonary artery arising from the posterolateral aspect of common arterial trunk (Truncus) from its posterolateral aspect and continuing as right pulmonary artery supplying right hilum.

The left major coronary artery exhibits an abnormal origin from the proximal aspect of the right pulmonary artery, which further divided into left anterior descending artery and left circumflex artery **(Figures 7 and 8)**. Right coronary artery has got its origin directly from common truncus.

Associated pulmonary artery hypertension with mosaic attenuation in bilateral lung fields were reported (**Figure 5**). Right heart strain was indicated by a contrast-related reflux from the right atrium into the inferior vena cava and hepatic veins.

Our patient showed truncus arteriosus with only RPA supplying right lung and absent MPA and LPA. A final diagnosis of type A3 truncus arteriosus (Van Praagh classification) was made. It is defined when single pulmonary artery that originates from the common trunk supplying one lung, while the other lung is fed by collateral arteries or a pulmonary artery emerging from the aortic arch[5]. {Video uploaded &VRT images-1& 2}

The modalities of the treatment, including the need for cardiac transplantation, were discussed with the family. However, the patient denied surgery and has been on regular follow-up for 1 year and has shown no worsening of symptoms.

Discussion

Persistent truncus arteriosus is a rare cyanotic congenital heart disease which is defined when single arterial trunk arises form the ventricle and is accompanied by large ventricular septal defect. While constituting fewer than 1% of the total number of congenital heart defects, the annual incidence is 7 per 100,000 live births and constitutes for 4% of severe congenital heart diseases[6].

The two main classification schemes for the truncus arteriosus are the Van Praagh & Van Praagh classification (1965) and the Collett and Edwards classification (1949).

Collett and Edwards classification[7]

Type 1: The main pulmonary artery and the aorta originate from the same trunk. (48–68% of cases)

Type 2 : Type 2 pulmonary arteries emerge from the posterior side of the trunk near to one another, directly above the truncal valve. (29–48% cases)

Type 3 : Pulmonary arteries emerge separately from each side of the trunk.(6–10% of cases)

Type 4 : Neither pulmonary arterial branch arising from the common trunk (pseudo truncus), considered a form of pulmonary atresia with a VSD

Van Praagh modified classification[5]

Type A1: Similar to Collett and Edwards' Type I, the aorta and primary pulmonary artery emerge from a similar trunk.

Type A2 : Both pulmonary arteries from the common trunk's left and right lateral aspects.

Type A 3 :Origin of single branch pulmonary artery from the common trunk, with other lung supplied by collaterals or a pulmonary artery arising from the aortic arch

Type A 4 : Coexistence of an interrupted aortic arch

In younger age groups, surgical surgery entails total correction through VSD closure and pulmonary artery to right ventricle connection[2]. Patients who have acquired pulmonary hypertension may undergo shunt procedure as palliative approach or dilation, may or may not involve the MAPCA stenting. Although there may be some benefit, the utility of medical management using phosphodiesterase-type 5 inhibitor sildenafil and endothelin antagonist is still not clear[8].

In our case, truncus arteriosus with single pulmonary artery (large aorta type with the absence of other pulmonary artery) falls under Type A3 variant. Desaturated blood is expelled into the single outflow tract due to mixing of pulmonary circulation blood and systemic circulation at VSD level. Major aortopulmonary collaterals offer high pressure pulmonary flow, which contributes to pulmonary arterial hypertension since the right pulmonary arteries directly arise from the truncus arteriosus. Heart failure is brought on by increased burden on the heart and lungs due to altered perfusion and ventilation mismatches. Larger MAPCAs and non-obstructive pulmonary arteries were the natural phenomenon for masking patient symptoms. Associated anomalous coronary artery and PAH have rendered this patient inoperative.

Embryologically, these lesions result from anomalies in the left sixth arch and cono-truncal anomalies. DiGeorge syndrome is typically linked to it[9]; however, a genetic investigation of our case has been planned.

After reviewing the literature, to the best of our knowledge, Truncus arteriosus with this kind of modified Van Praagh's type 3A surviving up to 26 years of age without need for surgery has not been documented in the literature till date. The survival of our patient offers information on the natural progression of the uncommonly complex congenital heart disease. The oldest similar reported case of this type of truncus was a 12-year-old child who survived without surgery[10].

Conclusion

Truncus arteriosus is a rare complex congenital heart disease, among which the Type A3 variant is one of the rarest[11]. In general, patients with truncus arteriosus exhibit symptoms during infancy and if not operated upon, their mortality rate is significantly high especially high in type A3 variant. Rarely do they grow to adulthood without receiving treatment. In other kinds of truncus arteriosus (other than A3 type), patients have been documented to reach adolescence and adulthood without receiving any therapy[12].

Our case, the patient with type A3 variant has survived till 26 years of age without any surgical intervention which makes it worthy of reporting.

Our case is one of the rare cases where a man of this variant of truncus has made it into his 3rd decade without surgery.

ABBREVIATIONS AND ACRONYMS

CE : Collett and Edwards
LPA : Left pulmonary artery
MPA : Main pulmonary artery
RPA : Right Pulmonary Artery
RV : Right ventricle
TA : Truncus arteriosus
VP :Van Praagh
VSD :Ventricular septal defect
LCX. : Left circumflex artery
LAD : Left anterior descending artery
CHD : Congenital heart disease
RCA : Right coronary artery

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