

## World Cardiology Summit 2020: De Bukes Syndrome - Tetralogy of Fallot with Absent Left Pulmonary Artery

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### Introduction:

De Buckes syndrome – TOF with unilateral absence of a pulmonary artery (UAPA) is a uncommon situation with an estimated incidence of 1 in 200,000 younger adults. Most commonly, UAPA takes place in conjunction with cardiovascular abnormalities such as tetralogy of Fallot (TOF) coarctation of the aorta, VSD, subvalvular aortic stenosis, transposition of the superb arteries (plus VSD or pulmonary stenosis), Taussig-Bing malformation and coarctation, congenitally corrected transposition and pulmonary stenosis, scimitar syndrome. Patients with remoted UAPA can stay asymptomatic into late maturity however commonly file signs and symptoms such as dyspnea or chest ache or go through from hemoptysis or recurrent infections. Diagnosis can be hard due to the rarity of the circumstance and its nonspecific presentation. We current a case of a 5month historic baby who introduced with TOF with Right pulmonary artery stenosis and absent left pulmonary artery, with standard findings on chest radiograph, angiographic elements and therapy discussed.

### Case Report :

A 5 months ancient male baby was once referred to Sri Jayadeva Institute of Cardiovascular Science and lookup for cardiac evaluation. He was once born to non-consanguinous mother and father with ordinary being pregnant and ordinary delivery. He introduced with records of incessant feeding and cyanosis whilst feeding and crying. On bodily examination, the affected person had cyanosis with SpO<sub>2</sub> of 67%. There was once no respiratory misery at rest. His weight used to be three kgs (just beneath the fifth percentile for his age). Heart price was once 104 beats/minute and blood stress used to be 100/55 mmHg. Cardiovascular examination published everyday peripheral pulses, the 2d coronary heart sound was once single and Grade 3/6 Ejection systolic murmur. The electrocardiogram confirmed sinus rhythm, proper axis deviation and proper ventricular (RV) hypertrophy. Chest X-ray confirmed moderate cardiomegaly, with regular bronchovascular markings on proper facet and absent bronchovascular markings on left side. A 2-dimensional echocardiography and Doppler learn about demonstrated the diagnoses of TOF with proper pulmonary artery stenosis and non-vislisation of left pulmonary artery, height systolic gradient of 70 mmHg throughout the RV outflow tract (RVOT). The aortic arch used to be left sided and no patent ductus arteriosus used to be seen. At cardiac catheterization, RV systolic stress was once at systemic stage and there used to be a height systolic gradient of seventy seven mmHg throughout RVOT. The principal PA strain was once 23/10 mmHg. The pulmonary drift to systemic drift (Qp/Qs) was once 0.83. The catheter should no longer enter the web page of LPA. PA angiography failed to exhibit a LPA. Angiography following pulmonary vein wedge injection in the left lung failed to exhibit LPA, confirming its absence. Aortography failed to exhibit a patent ductus arteriosus, collaterals presenting the distal phase of LPA or anomalous starting place of the LPA. The analysis of TOF with proper pulmonary artery stenosis and absent LPA was once established. Since

the baby was once symptomatic, pulmonary artery balloon dilatation of proper pulmonary artery used to be carried out which reduced the gradient throughout the RVOT with temporary alleviation of symptoms. The baby used to be later referred to cardiothoracic healthcare professional for definitive intracardiac repair.

### Discussion :

UAPA is a uncommon condition, with an estimated incidence of 1 in 200,000 younger adults. Most commonly, UAPA happens in conjunction with cardiovascular abnormalities such as tetralogy of Fallot or cardiac septal defects, however it can additionally take place in an remoted manner. Isolated UAPA entails the proper lung in about two thirds of cases. Due to embryologic relationships, UAPA usually takes place on the aspect of the chest contrary the aortic arch (although that was once no longer the case in our patient). The actual embryologic purpose of UAPA is a count of debate and is probably specific in left- vs. right-sided UAPA. In each cases, however, altered improvement of a sixth aortic arch phase is concept to end result in a ductal beginning to a pulmonary artery that leads to the proximal interruption of that vessel when the ductal tissue regresses at the time of birth. Distal intrapulmonary branches of the affected artery commonly continue to be intact and can be provided by way of collateral vessels from bronchial, intercostal, inner mammary, subdiaphragmatic, subclavian, or even coronary arteries. Patients with remoted UAPA can current in a range of ways. A 2002 assessment of 108 instances of UAPA published a median age of presentation of 14 years. The aggregate of chest pain, pleural effusion, and recurrent infections was once existing in 37% of patients, whilst dyspnea or exercising intolerance was once current in 40% of patients. Pulmonary hypertension used to be observed in 44% of sufferers that had been examined for the disorder. Hemoptysis came about in about 20% of patients, and high-altitude pulmonary edema used to be viewed in about 10% of patients. Seven deaths had been stated in the case collection and covered mortality from big pulmonary hemorrhage, proper coronary heart failure, respiratory failure, pulmonary hypertension, and high-altitude pulmonary edema. Only 14 of 108 sufferers with remoted UAPA have been asymptomatic at the time of their analysis and all through variable followup. In addition, negative blood waft to the affected lung might also end result in alveolar hypoxemia, main to secondary bronchoconstriction and mucous trapping. Chronic contamination can lead to bronchiectasis in some patients. Hemoptysis is a doubtlessly serious complication of UAPA. Hemoptysis seems to be brought on by way of giant collateral circulations that difficulty venous structures to surprisingly excessive pressures. While hemoptysis can be continual and self-limited, instances of huge hemoptysis have been mentioned in the literature.

Diagnosing UAPA can be difficult, however necessary clues are current in chest radiographs. The chest radiograph of sufferers with UAPA normally suggests uneven lung fields, with an ipsilateral small hemithorax protecting a hyperlucent lung. When suspicious findings are stated on a chest radiograph, the analysis of UAPA can be definitively made via CT, magnetic resonance imaging (MRI), or transthoracic echocardiogram. On cross-sectional imaging, the absent pulmonary artery will normally terminate inside 1 cm of its anticipated starting place from the predominant pulmonary artery. Other findings that may additionally be referred to on CT or MRI encompass intact peripheral branches of the pulmonary artery, variable collateral circulation, mosaic parenchymal changes, and bronchiectasis secondary to recurrent infections.

Transthoracic echocardiogram can additionally be used to diagnose UAPA and is tremendous due to the fact the examiner can seem to be for coexisting cardiac malformations at the identical time. Angiography is viewed the gold popular for the analysis of UAPA however is invasive and usually unnecessary, until it is being used as a preoperative check for a reading hemodynamic parameters and additionally for palliative treatment. There is presently no consensus regarding therapy of sufferers with UAPA. Some authors have endorsed the use of serial echocardiography to display asymptomatic adults for the improvement of pulmonary

hypertension. Patients who enhance pulmonary hypertension can be dealt with medically with vasodilator therapy. Alternatively, revascularization of peripheral branches of the affected pulmonary artery to the pulmonary hilum can be attempted, and there are reviews of profitable revascularization procedures, usually in the pediatric population.

**Conclusion:**

Agenesis of the left pulmonary artery is a uncommon anomaly and the affected person may additionally continue to be asymptomatic till adulthood. Imaging performs a important function in the prognosis and detecting the related findings in coronary heart and lungs. For sufferers who current with a unilateral small hemithorax, recurrent chest infections and an strange chest X-ray, a UAPA anomaly need to be regarded in the differential diagnosis. An early analysis can forestall serious complications.