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Immediate Surgical Outcome of Obstructed Infracardiac Total Anomalous Pulmonary Venous Return with Unilateral Renal Cyctic Dysplasia: Case Report

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Abstract

Obstructed infracardiac total anomalous pulmonary venous return (TAPVR) is one of the cyanotic congenital heart disease requiring urgent surgical intervention. Surgical repair of obstructed infracardiac TAPVR with severe pulmonary hypertension is still a challenge in low economic settings due to lack of advanced care for the management of anticipated severe pulmonary hypertension and left ventricular dysfunction. We report a case of a neonate admitted for evaluation due to prenatal ultrasound findings of cystic right kidney but later developed increasing respiratory distress on second day of life. A diagnosis of obstructed infracardiac TAPVR with severe pulmonary hypertension was made by echocardiography and CT angiography. He underwent successful TAPVR repair as the first case of obstructed TAPVR to undergo surgical repair at Jakaya Kikwete Cardiac Institute (JKCI) with good immediate outcome.

Keywords: Total anomalous pulmonary venous return (TAPVR); Obstructed infracardiac TAPVR; Renal cysts; Immediate outcome

Introduction

Total anomalous pulmonary venous return (TAPVR) is a cyanotic congenital defect in which all four pulmonary veins fail to make their normal connection to the left atrium resulting into drainage of all pulmonary venous return into the systemic venous circulation. The incidence of TAPVR ranges from 0.6 to 1.2 per 10,000 live births [1]. There are four anatomical types depending on the level of anomalous venous drainage: supra cardiac (50%), cardiac (15%), infra cardiac (20%) and mixed type (10%) [2]. In patients with TAPVR, the entire oxygenated pulmonary venous return mixes with deoxygenated blood from the systemic venous system resulting into cyanosis.

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It has variable clinical presentation depending on the type and degree of pulmonary venous obstruction thus requiring high index of suspicion to diagnose. Obstruction may occur with all types but is commonest with infracardiac type, where it may occur, when the vertical vein crosses the diaphragm or at point of connection with the hepatic, portal veins or the inferior vena cava (IVC). Obstructed TAPVR presents with severe respiratory distress due to pulmonary congestion and pulmonary hypertension, increasing the risk of preoperative morbidity and mortality [3]. Early intervention for obstructed TAPVR is usually required for survival. In severe cases of obstructed TAPVR with severe pulmonary hypertension, advanced care with extracorporeal membrane oxygenation (ECMO) or nitric oxide increases survival [4]. TAPVR usually occurs as an isolated cardiac lesion but it can occur with other congenital anomalies such as in heterotaxy with polyspenia or asplenia syndromes [3,5]. Risk factors for postoperative mortality include younger age at surgery, associated cardiac lesions, hypo plastic pulmonary veins, postoperative pulmonary hypertension and postoperative pulmonary venous obstruction [6].

We report a case of obstructed infracardiac TAPVR with severe pulmonary hypertension and right kidney cysts.

Case Presentation

A term non-syndromic, male baby weighing 3.6 kg was delivered by Caesarian section due to previous scar scoring 8 and 10 at first and fifth minutes respectively. Admission at Muhimbili National Hospital neonatal ward was done for evaluation due to findings from a routine prenatal ultrasound in 3rd trimester that showed abnormal right kidney. Mother was 26 years old and had uneventful pregnancy. On 2nd day of life, baby developed fast breathing and inability to breastfeed well. On exam, he had tachypnea, respiratory rate of 70 breaths /minute and oxygen saturations of 90% room air. He was kept on oxygen by nasal prongs 1 litre/minute and presumptive treatment for early neonatal sepsis. On 3rd day, respiratory distress worsened with lower saturations (80%-85%) associated with severe respiratory acidosis. Septic markers and renal function tests were within normal limits. Chest x-ray showed bilateral homogenous haziness suggestive of pulmonary congestion. Echocardiography demonstrated total anomalous pulmonary venous return into a common confluence that descended down via a vertical vein to the hepatic veins then into inferior venacava (IVC) which then drained into right atrium (Figure 1). Tricuspid regurgitation (TR) maximal velocity was 5m/sec indicating severe pulmonary hypertension. Obstruction was noted at the point where vertical vein joined the hepatic veins (maximal velocity 3m/s) and at the IVC entry into right atrium (maximal velocity 3m/s). Renal ultrasound and CT scan showed multiple cysts in the right kidney and a normal left kidney (Figure 2). Cardiac CT scan demonstrated all pulmonary veins draining into a common venous channel descending through the esophageal hiatus and connects to main portal vein and left hepatic vein (Figure 3). He was kept on mechanical ventilation on maximum settings and infusion Furosemide 0.2mg/kg/hour for pulmonary edema. Patient was transferred to Jakaya Kikwete Cardiac Institute (JKCI) for obstructed TAPVR repair. This was the first case of obstructed infracardiac TAPVC to undergo complete repair at JKCI. In view of the patient's hemodynamic instability, and severe pulmonary hypertension in resource-limited settings relying only on sildenafil with the lack of advanced care for severe pulmonary hypertensive crisis such as nitric oxide and Extracorporeal Membrane Oxygenation (ECMO); parents were counselled for a high-risk surgery.

Complete repair was done on day 11 of life under deep hypothermia and circulatory arrest via a "suture less" technique where the left atrium was sutured to the posterior pericardial wall around the common pulmonary venous chamber. A 4mm patent ductus arteriosus (PDA) was ligated and the large secundum atrial septal defect (ASD) was downsized to a smaller ASD as a "pop off" shunt in the event of pulmonary hypertensive crisis.

Cardiopulmonary bypass time was 114 minutes with no major intraoperative event. Chest was left open to allow the edematous heart and lung to subside. Delayed chest closure was done 48 hours postoperatively without incident. Immediate post-operative echocardiogram showed unobstructed pulmonary venous drainage into the left atrium, trace tricuspid regurgitation and good biventricular function.

On day three postoperatively; patient experienced episodes of pulmonary hypertensive crisis associated with apnea, bradycardia and hypotension that was managed by cardiopulmonary rescuscitation and inotropic support (Milrinone). Other post-operative issue was residual pulmonary edema that was managed by intensified diuresis and chest physiotherapy.

Patient was weaned from mechanical ventilation and inotropic support on postoperative day 13 and 14, respectively. Total ICU stay was 20 days and total hospital stay was 46 days. Follow up was done at 2 weeks, 1 month, 3 months and at 6 months post discharge he had a normal physical examination, good results on postoperative echocardiography and normal renal functions.

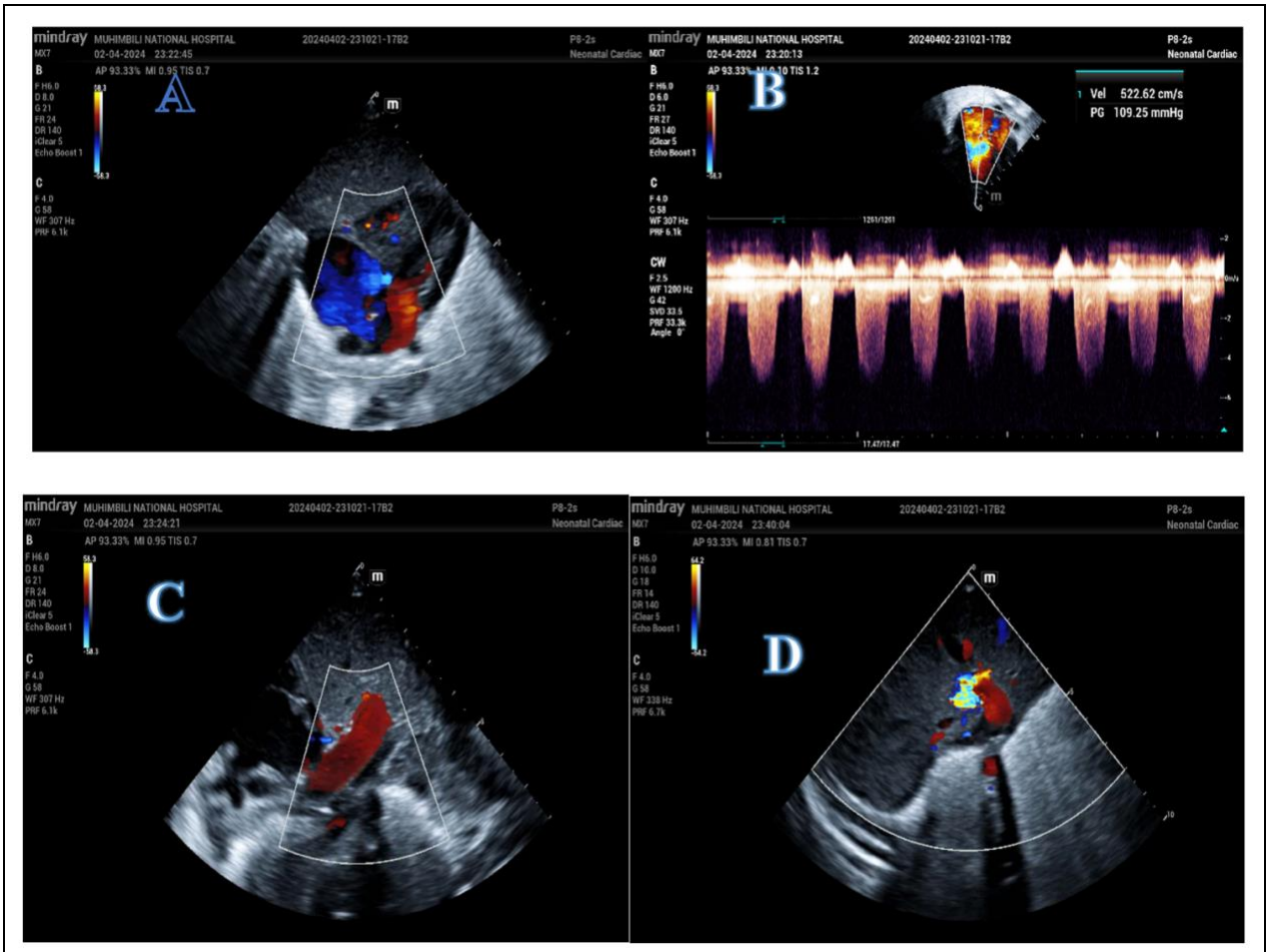


Figure 1: Color Doppler echo subxiphoid view; (A) showing a dilated right atrium, underfilled left atrium and right to left shunting through an ASD secundum into the left atrium. Tricuspid regurgitation jet of velocity 5.22m/s indicating severe pulmonary hypertension (B). Color Doppler echo showing the descending vertical vein entering into the liver to join the hepatic veins (C,D). A mosaic pattern noted as vertical vein joins the hepatic veins signifying obstruction at point of entry (D).

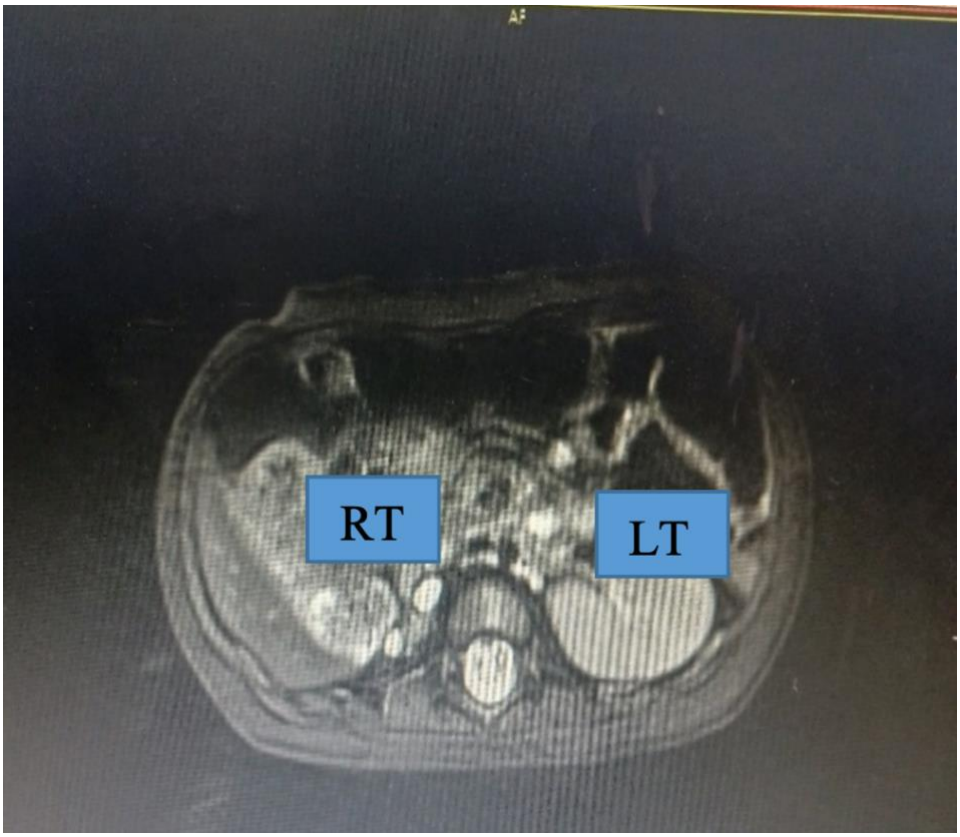


Figure 2: Renal MRI-T2W axial view-poor contrast uptake in Right (RT) kidney, two small cysts. Good contrast uptake is seen in the normal left (LT) kidney.

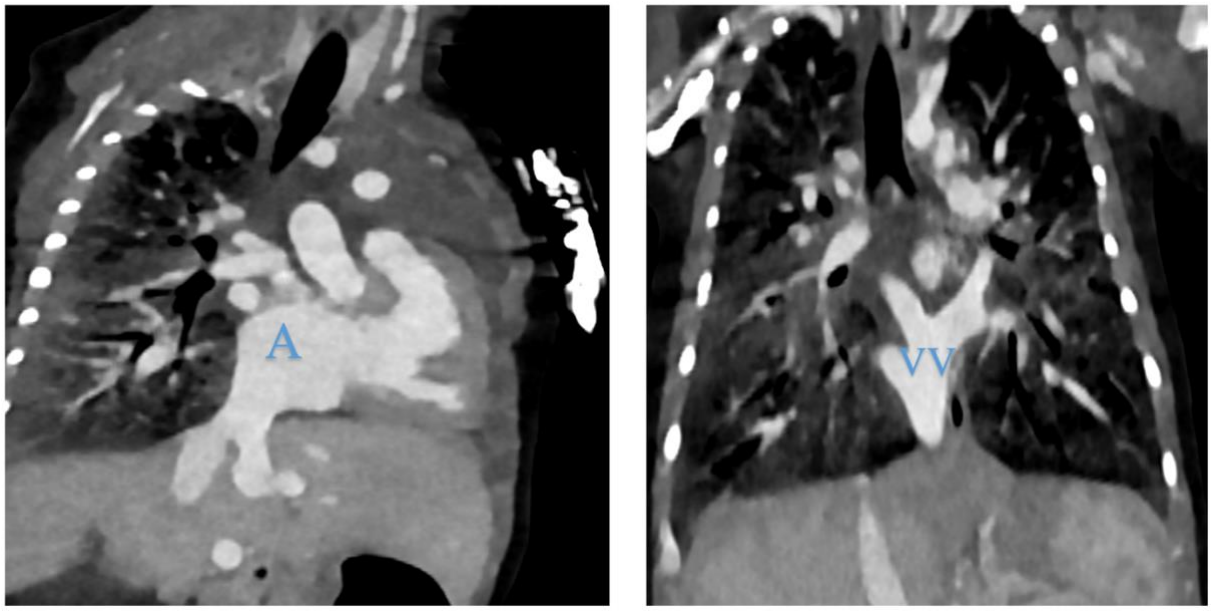


Figure 3: CT scan angiography in coronal view showing four pulmonary veins collecting into a common confluence (A) then descending down through a vertical vein (VV) through the diaphragm to join the hepatic veins.

Discussion

Obstructed infracardiac total anomalous pulmonary venous return (TAPVR) is one of the cyanotic congenital heart defects that present during the neonatal period requiring urgent intervention for survival. Obstructed infracardiac TAPVR associated with severe pulmonary hypertension carries increased risk for both pre- and post-operative mortality [7].

We presented a case of a neonate with obstructed infracardiac TAPVR associated with a severe pulmonary hypertension and a dysplatsic right kidney, who underwent TAPVR repair as the first case of its kind at Jakaya Kikwete Cardiac Institute (JKCI) with good immediate surgical outcome. Repair of obstructed infracardiac TAPVR is still one of the challenging procedures especially in limited resourced areas that lack advanced care such as nitric oxide to manage cases of severe pulmonary hypertensive crises. Presence of severe pulmonary hypertension as in this case increases risk of pre- and post-operative mortality [7] especially in settings with limited options for managing anticipated pulmonary hypertensive crisis.

Other factors that have been reported to increase mortality in obstructed TAPVR are coexisting cardiac anomalies such as single-ventricle physiology, younger age at operation, need for emergency repair, increased total bypass and circulatory arrest time and pulmonary venous obstruction (PVO) [6,8]. This patient had right kidney cysts suggestive of a dysplastic right kidney disease. Although no reported familial history of congenital kidney or heart disease, several reports have shown common coexistence of congenital heart diseases with congenital kidney anomalies by 30% and a shared genetic etiology is involved [9]. The unilateral involvement of the right kidney and a normal left kidney predicts good prognosis due to higher chances of spontaneous involution of the dysplastic kidney but patient requires long-term renal follow-up due to reported risk of developing cystic infection, hypertension, renal failure or malignancy later in life [10].

In the period of 6 months that the patient was followed up, echocardiography demonstrated no evidence of pulmonary venous obstruction but long-term follow up is required for pulmonary vein stenosis. It has been reported that; preoperative pulmonary vein obstruction increases risk for recurrent pulmonary obstruction after surgery, and thus increased risk of both early and late morbidity and mortality [11]. This case used the “suture less” surgical technique for TAPVR repair as it has shown to reduce the risk for postoperative pulmonary vein stenosis [12].

Surgical outcome and survival of isolated TAPVR has improved due to advancement of medical and surgical management, and the use of extracorporeal membrane oxygenation (ECMO) to support the most severely affected neonates [13]. The early mortality (<30 days) is reported to be less than 10% [7,14] in well-developed centres.

This case had uneventful surgery but developed postoperative pulmonary hypertensive crisis and residual pulmonary congestion that were managed despite having limited resources. Although lack of advanced care limits optimal post-operative care, this case has built foundation for surgical repair of other subsequent similar cases in our setting. Long-term follow-up of this case is required for assessing pulmonary veins stenosis and evaluation of the kidneys.

Conclusion

Immediate surgical outcome of obstructed infracardiac TAPVR with severe pulmonary hypertension in resource-limited institution is promising. To ensure good outcome, we recommend advancing cardiac care in order to optimize care of postoperative patients with pulmonary hypertensive crisis. Coexistence of congenital renal and cardiac malformations is not uncommon thus, early screening is important.

Acknowledgement

We would like to acknowledge the parents of this child who agreed to give the required information and consent for publication of this case.

Declarations

Ethical approval: Ethical approval was obtained from the research and publication committees of Muhimbili National Hospital and Jakaya Kikwete cardiac institute.

Competing interest: Authors declares that they have no competing interest and this case was not funded.

Authors contributions: YK was involved in writing the case and preparation of the final manuscript. DN, SM, GS, VM and SK critically reviewed the case.

All authors approved the final manuscript for publication.

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