Difficult vascular access for hemodialysis in congenital bilateral absence of radial artery and probable VACTERL association: a case report

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Abstract

Isolated bilateral absence of radial artery in association with other congenital anomalies, together named VACTERL (vertebral defects, anal atresia, cardiac defects, tracheo-esophageal fistula, renal anomalies, and limb abnormalities) is a very rare phenomenon. We report a patient, known to have VACTERL association, and who presented with ESRD. He was on hemodialysis and had been subjected to multiple failed tries at vascular access due to congenital bilateral absence of radial arteries and other vascular anomalies. This case report highlights hemodialysis options in congenital bilateral absence of radial artery and probable VACTERL association showing superiority of arteriovenous fistula despite its accompanying complications.

Key words: VACTERL; Abnormalities, Multiple / classification; Kidney / abnormalities; Limb Deformities, Congenital; Bilateral absent radial artery; Hemodialysis; AV fistula, Failure; AV fistula, Complications

Abbreviations: VACTERL - Vertebral defects, Anal atresia, Cardiac defects, Tracheo-esophageal fistula, Renal anomalies, and Limb abnormalities; AVF - Arteriovenous fistula; ESRD - End-stage renal disease; AVG - Arteriovenous graft; CVC - Central venous catheter


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1. Introduction

Blood supply to the forearm and hand is mainly by two branches of the brachial artery: radial and ulnar arteries.1 These arteries, especially the radial arteries, are preferred access sites for many interventions including percutaneous coronary intervention2 and arteriovenous fistula (AVF) formation for hemodialysis.3 Congenital anomalies of the radial artery are very uncommon and most of them are related to the site of origin of the artery or its course.4,5 Complete absence of radial artery is very rare with only twelve reported cases in the literature; eight of these were of unilateral aplasia and only four cases reported bilateral absence of the artery without any associated vascular or non-vascular malformations.6,7

This paper presents a case of a female patient with end-stage renal disease (ESRD) and bilateral absence of radial arteries that were discovered after AVF failure. The case was a complex of multiple defects that give a probable picture of VACTERL association; which is a sporadic multifactorial association of non-random multisystem birth defects comprising vertebral defects, anal atresia, cardiac defects, tracheo-esophageal abnormalities, renal anomalies and limb defects as the acronym VACTERL implies.8,9

There are three types of vascular hemodialysis access: AVF, arteriovenous graft (AVG) and central venous catheter (CVC); each method has its pros and cons, but overall, the purpose is to create an access point with good blood flow, ability to withstand repeated needle
insertions and with the lowest possible risk of complications. We discuss the complexity of vascular access for hemodialysis in a setting of multiple vascular anomalies in this case report.

2. Case Report

A 42-year-old female patient, known case of ESRD, on hemodialysis, had multiple failed vascular access points including AVF and temporary and long-term CVC insertion. The condition started about twelve years back with declining renal function, ultrasound of the abdomen showed agenesis of the right kidney and a small sized left kidney with increased parenchymal echogenicity and loss of corticomedullary differentiation (not enough data to suggest the inciting event that led to the pathological picture of the left kidney). Physical examination of the patient showed miniature body habitus (height 145 cm, weight 39 kg), and congenital hypoplastic thumb on both sides (thumb on the right side was surgically removed) (Figure 1). The patient’s family reported a history of imperforate anus that was corrected surgically. She had also suffered pain and stiffness in her neck and x-ray showed segmentation, deformity, fusion and partial agenesis of multiple cervical vertebrae (Figure 2). This multisystem involvement with no associated family history suggested a probable diagnosis of VACTERL association.

Four years back, her first AVF was made in the right cubital fossa to prepare her for hemodialysis due to continuous decline in renal function. Surgical intervention was done without a preceding vascular mapping and the patient was lost to follow up afterwards. When she reported back in June 2019, the fistula had failed to mature. Duplex ultrasound of both upper and lower limbs was carried out to decide on the appropriate site for vascular access. Scan showed absence of both radial arteries as well as great saphenous veins on both sides; left cubital fossa had a brachial artery with a diameter of 2.5 mm, median cubital vein of 1.8 mm and cephalic vein of 2 mm, and all were patent with normal blood flow. So, a new fistula was created on the left side by an end to side anastomosis of the median cubital vein and brachial artery. During that period, a CVC was inserted in the right internal jugular vein to start hemodialysis, this was followed by insertion of catheters in femoral veins, each lasting for two or three weeks. After two months, the fistula was fully mature, and hemodialysis was started through it and persisted for about six months without complications. Then secondary fistula failure was noticed due to thrombosis which might be a complication of COVID-19 infection and a direct external trauma on the fistula site. A revision surgery was done to create an end to side anastomosis of the cephalic vein and brachial artery, the vein was ligated distally and isolated from the previous AVF before being anastomosed with the brachial artery. During that period, a CVC was inserted in the left subclavian vein to use as a port for hemodialysis while waiting for the AVF to mature. There were multiple sites used for catheter insertion.

Patient resumed hemodialysis through AVF after fourteen days and persisted for almost a year. After which, due to thrombosis and repeated cannulation, fistula was complicated by development of two aneurysms that necessitated reoperation; a big thrombosed aneurysm was resected and another one smaller and non-thrombosed was repaired by aneurysmorrhaphy (Figure 3). A new AVF was created by end to side anastomosis of the cephalic vein and brachial artery. A CVC in the right subclavian vein was used while waiting for maturation of AVF. Hemodialysis was restarted through the fistula after sixteen days and was still in use at the time of the last contact with the patient on 16th March 2022.

3. Discussion

A VACTERL patient can have the diagnosis apparent at birth, or only some features can be recognized in the neonatal period with the others remaining undetectable.
Screening tests should be considered in cases of multiple congenital anomalies without a clear identifiable cause to look for possible associated defects. In this case, anal and limb anomalies were apparent at birth and no screening was done to detect the other defects. Fortunately, she didn’t have cardiac involvement, which if present and undetected early, would lead to heart failure later in life. Detection of vascular anomalies were delayed until renal replacement therapy with hemodialysis was needed.

Choice of vascular access type for hemodialysis is a matter of debate, and studies in this field do not provide high quality evidence to support the use one against the others in all instances. However, the scientific community agrees that mature fistula is superior to AVG and CVC for long term hemodialysis due to lower risk of thrombosis, infection, hospitalization and all cause morbidity and mortality. But primary fistula failure remains an important and high-risk cause for morbidity and mortality with an estimated incidence of 30-50%. Many factors increase the risk of primary failure or lead to difficult or even impossible AVF creation, including vessel size, vascular disease, unfavorable anatomy and history of previous failed AVF, like in this reported case. An important factor that may had led to failure of the first AVF in this patient was the inadequate knowledge of vascular anatomy and the condition of blood flow. Therefore, vascular mapping and proper planning before creation of AVF can decrease the rate of failure and the need for unnecessary interventions. Despite superiority of AVF in terms of longevity, thrombosis can be a cause of secondary failure after full maturation and initiation of hemodialysis. In this case, the coincidence of thrombosis as the cause of AVF failure with COVID-19 infection and direct external trauma was unexpected and didn’t follow the classical association of thrombosis with intimal hyperplasia.

Aneurysm is another complication of established AVF that can be a consequence of weakened vessel wall due to repeated cannulation or the presence of outflow stenosis. With this in mind, it would be rational to try avoiding the placement of a CVC proximal to an AVF in patients who need another access point for any reason; as catheters can induce scarring and stenosis of veins. Furthermore, if a stenosis of the draining vein is detected when treating a fistula with aneurysm, an important step would be to consider proximal percutaneous transluminal angioplasty.

When AVF is not feasible, AVG is the second-best choice being even superior to AVF in terms of higher primary patency. In a study done by Uzun et al., saphenous vein graft was preferred over synthetic grafts in terms of higher primary and secondary patency, but the absence of both saphenous veins in this patient precluded this option. CVCs were used as a bridge when waiting for maturation of an AVF. Unfortunately, the longest duration a catheter lasted was around three to four weeks and all of them were complicated by thrombotic occlusion. In addition to the risk of infection especially when using a catheter.
in the femoral vein, all these factors make CVC a last choice and to be used only if absolutely necessary.

4. Conclusion

Vascular access longevity and the risk of complications determine the quality of life of patients on long term hemodialysis. Despite absence of both radial arteries that eliminate the possibility of distal AVF and recurrent complications in the form of thrombosis and aneurysm in established fistulae, AVF was the most suitable access for hemodialysis in this VACTERL patient.

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6. Conflict of interest

Authors declare no conflict of interest.

7. Patients’ consent

The patient agreed to allow the authors to publish her case details and images, and to provide upon request.

8. Authors contribution

GK: Drafting; conduction of the study work and manuscript editing

WM: Write-up; concept; and conduction of the study work

9. References


