# CHALLENGING CATHETER MANAGEMENT IN A RARE CASE OF MYELOMENINGOCELE WITH SITUS INVERSUS TOTALIS: A CASE REPORT

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## ABSTRACT

Individuals diagnosed with situs inversus totalis and renal failure have distinctive difficulties, such as an increased tendency to vascular stenosis and thrombosis during dialysis as a result of changed vascular patterns. A 19-year-old woman with myelomeningocele and situs inversus totalis is diagnosed with end-stage kidney disease (ESKD) as a result of bladder dysfunction. Due to the inadequacy of her vascular access, she commenced hemodialysis with the right permanent catheter. Regrettably, she developed a central line-associated bloodstream infection caused by Methicillin-Susceptible Staphylococcus aureus (MSSA), leading to septic shock. A CT scan and echocardiography revealed 9-mm tricuspid vegetation located around the cavoatrial junction. After a six-week course of intravenous cloxacillin, she was discharged with a left superior vena cava catheter. Without appropriate treatment, vascular issues might have serious repercussions for ESKD patients. Situs inversus presents extra challenges. Putting a catheter in a dextrocardia patient requires special skills. They also have unique vascular anatomy that necessitates different management than typical ESKD patients. Thus, a proper catheter size and type and imaging modalities like ultrasonography or venography are needed to simplify insertion and reduce complications. Renal failure treatment is perplexed by the presence of situs inversus totalis.

**Keywords:** Situs Inversus Totalis, Dextrocardia, ESKD, permanent catheter, vascular stenosis

## INTRODUCTION

Situs inversus totalis typically does not result in any significant harm to an individual; however, it can occur alongside other congenital abnormalities, including those affecting the cardiovascular system (ventricular septal defect, atrial septal defect, tetralogy of Fallot, transposition of great arteries), the respiratory system (bronchiectasis, paranasal sinus deformity), and the digestive system (such as anal atresia, duodenal atresia) (Vaid et al.,2015). Patients with situs inversus totalis may develop renal failure as a result of many issues, such as post infectious glomerulonephritis, IgA nephropathy, and cystic kidney disease. The occurrence of spinal dysraphism in conjunction with situs inversus totalis is extremely unusual, and to this day, only a handful of examples have been documented (Vaid et al.,2015). The commonest spina bifida is myelomeningocele. It can be associated with hydrocephalus, Arnold Chiari malformation, VACTERL (vertebral defects, anal atresia, cardiac defects, tracheo-esophageal fistula, renal anomalies, and limb abnormalities), and OEIS (Omphalocele-Exstrophy-Imperforate Anus-Spinal Defects). This case report delves into the complex medical journey of a 19-year-old girl who faced a multitude of complications stemming from this rare combination.

## CASE REPORT

A 19-year-old girl with myelomeningocele and situs inversus totalis caused complications include leg paralysis, bladder dysfunction, scoliosis, hydrocephalus, and cognitive impairment. Shunt placement for hydrocephalus and scoliosis correction were needed for the patient. Her neurogenic bladder caused vesicoureteral reflux and recurring urinary tract infections, resulting in ESKD. Due to her ESKD, the patient started hemodialysis, however her small size prevented fistula development. Dextrocardia rendered catheter insertion difficult and uncomfortable for the patient. The patient had repeated catheter exchanges due to catheter malfunction after catheter implantation.

After three months on a permanent catheter, the patient came to the HDU for dialysis session with fever, cough, and shortness of breath intradialysis. The patient was not in respiratory distress, and the tunnelled catheter exit site discharged little pus. A CT scan revealed situs inversus, big impacted faeces in the rectum, focal fat stranding in the right paracolic gutter in the omentum, mild bilateral hydroureteronephrosis with diffusely enhanced urothelium, and several osseous lytic lesions, most likely brown tumours.

Based on the patient's clinical presentation, the patient was diagnosed with septic shock with a possible CLABSI (central line bloodstream infection). Urgent recommendations were made to arrange for a three-hour HD session through the available tunnelled catheter, remove it immediately after, and keep the patient in line-holiday for 24-48 hours. Additionally, arranging for Quinton line insertion through femoral vein for the next session.

Despite being on long-term antibiotics, the patient did not show signs of recovery, and echocardiogram was done, revealing a 9-mm multilobulated hypodense lesion attached to the tricuspid valve posterior/septal leaflet, likely representing vegetation. Furthermore, an ill-defined hypodensity at the cavoatrial junction was causing mild luminal narrowing, likely representing thrombus formation. Bilateral subpleural consolidative densities with areas of central lucency were highly suggestive of septic emboli. Her blood culture shows methicillin-sensitive Staphylococcus aureus and requires a 6-week antibiotic course. Repeated ECHO shows resolution of tricuspid vegetation. She was temporarily dialyzed via a femoral catheter during her hospital stay. She was discharged well at the opposite site (Left) of permanent catheter insertion and is awaiting a vascular appointment for fistula creation.

## DISCUSSION

This case illustrates the complex history of a 19-year-old girl with several congenital abnormalities and multiple complications requiring ongoing medical management and surgical interventions. She has myelomeningocele, which is a subset of spina bifida. Spina bifida, which is the most common congenital abnormality of the central nervous system and involves a problem with neural tube closure. It may be classified anatomically into open spina bifida and closed spina bifida, with the former including myelomeningocele and myelocele as the most common forms.

A spinal cord segment protruding through a backbone gap causes myelomeningocele, which causes neurological impairments and other issues (Ntimbani et al., 2019). Myelomeningocele can induce limb muscle weakness or paralysis, bowel and bladder dysfunction, scoliosis, and cognitive deficits, requiring constant medical management and a multidisciplinary approach. Damage to the nerves that regulate the bladder muscles and sphincters can cause urine incontinence, UTIs, and other issues. Additionally, urinary tract infections, vesicoureteral reflux, and obstructive uropathy can cause end-stage kidney disease. Treatment aims to improve function and quality of life while reducing consequences and medical issues, including emotional and social challenges. Conventional postnatal surgical repair of a myelomeningocele defect is performed as soon as practically feasible after the exclusion of life-threatening associations, ideally during the first 48 hours of birth, to limit the danger of infection, chronic cerebrospinal fluid leakage, and for convenience of care.

Besides that, she has situs inversus, which is a rare genetic condition in which the organs in a person's chest and abdomen develop in a right-left formation instead of the normal left-right formation. Situs inversus can be associated with cystic kidney diseases and chronic renal failure. However, only three genes have been found to be responsible for some cases of cystic kidney diseases in situs inversus patients (Onoe et al., 2013). Patients with situs inversus who have chronic renal failure may have symptoms such as pallor and other signs of renal insufficiency (Mittal et al., 1994). Imaging tests such as an ultrasound, X-ray, CT scan, or MRI scan can be used to diagnose situs inversus and chronic kidney disease. If complications related to another defect or condition occur, these can be treated. It is important to inform healthcare providers if you have situs inversus so that future ailments can be diagnosed accurately due to the mirrored anatomy.

On the other hand, there are few types of dextrocardia that can be identified, which include dextrocardia with situs solitus, with situs inversus, and with situs ambiguous. Mirror-image Dextrocardia with situs inversus, inverted great arteries, and L-loop ventricles is the result of situs inversus with concordant L-bulboventricular loop, which is the opposite of normal cardiac anatomy (Maldjian et al., 2007). Dextrocardia occurs due to the migration of the L-bulboventricular loop's apex into the right hemithorax, which is the most common type of dextrocardia in the general population, occurring in one or two out of every 20,000 individuals .

In dextrocardia, the positions of the vessels that enter and exit the heart are reversed, with the aorta arising from the right ventricle and the pulmonary artery arising from the left ventricle, contrary to the usual positioning. This anomaly also extends to the superior vena cava, located left of the midline, and the inferior vena cava, situated to the right of the midline, in contrast to their normal anatomical positions (Maldjian et al., 2007). Furthermore, the coronary arteries responsible for supplying blood to the heart muscle are reversed in dextrocardia, with the right coronary artery arising from the left side of the aorta and the left coronary artery arising from the right side of the aorta, which differs from typical

cardiac anatomy. These unique anatomical differences in dextrocardia hold significant implications for the diagnosis and medical treatment of affected individuals. Due to this condition, catheter insertion is more difficult because most catheters are designed for people with normal anatomy. Guided by the KDOQI 2020 recommendations, catheter insertion should prioritize vascular access sites in the following order: right internal jugular, left internal jugular, external jugular, femoral, subclavian, and lumbar veins, to minimize procedural complications (Lok et al., 2019). However, in cases of situs inversus, where anatomy is mirrored, the preferred site for catheter placement should be the left internal jugular vein to accommodate the reversed anatomical landmarks.

Insertion of a catheter in a dextrocardia patient can be challenging, and multiple attempts may be necessary before successful placement. However, multiple attempts can also increase the risk of complications such as bleeding, infection, and damage to surrounding structures. To minimise the risk of complications during catheter insertion, it's important to implement catheter care bundle standard of procedure, select an appropriate catheter size and type, and ensure proper placement and securement of the catheter. Imaging plays a crucial role in identifying vessels for catheter insertion in patients with dextrocardia who require hemodialysis. Imaging techniques such as ultrasound or venography can help locate suitable vessels for catheter insertion and guide the placement of the catheter. Additionally, this can help to minimise the risk of complications and improve patient outcomes.

Furthermore, the synergistic collaboration amongst the nephrology, vascular, and interventional radiology teams is paramount. A unified approach and ongoing dialogue within this multidisciplinary team, aligned with the principles of the 'End-Stage Kidney Disease Life Plan,' serve not merely to preclude dialysis access-related complications but also to enhance overall patient morbidity and mortality outcomes. This holistic strategy underscores the necessity of a coordinated care paradigm, ensuring that each catheter insertion is not only technically successful but also aligned with the patient's broader treatment objectives and quality of life considerations.

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**FIGURE LEGENDS:**

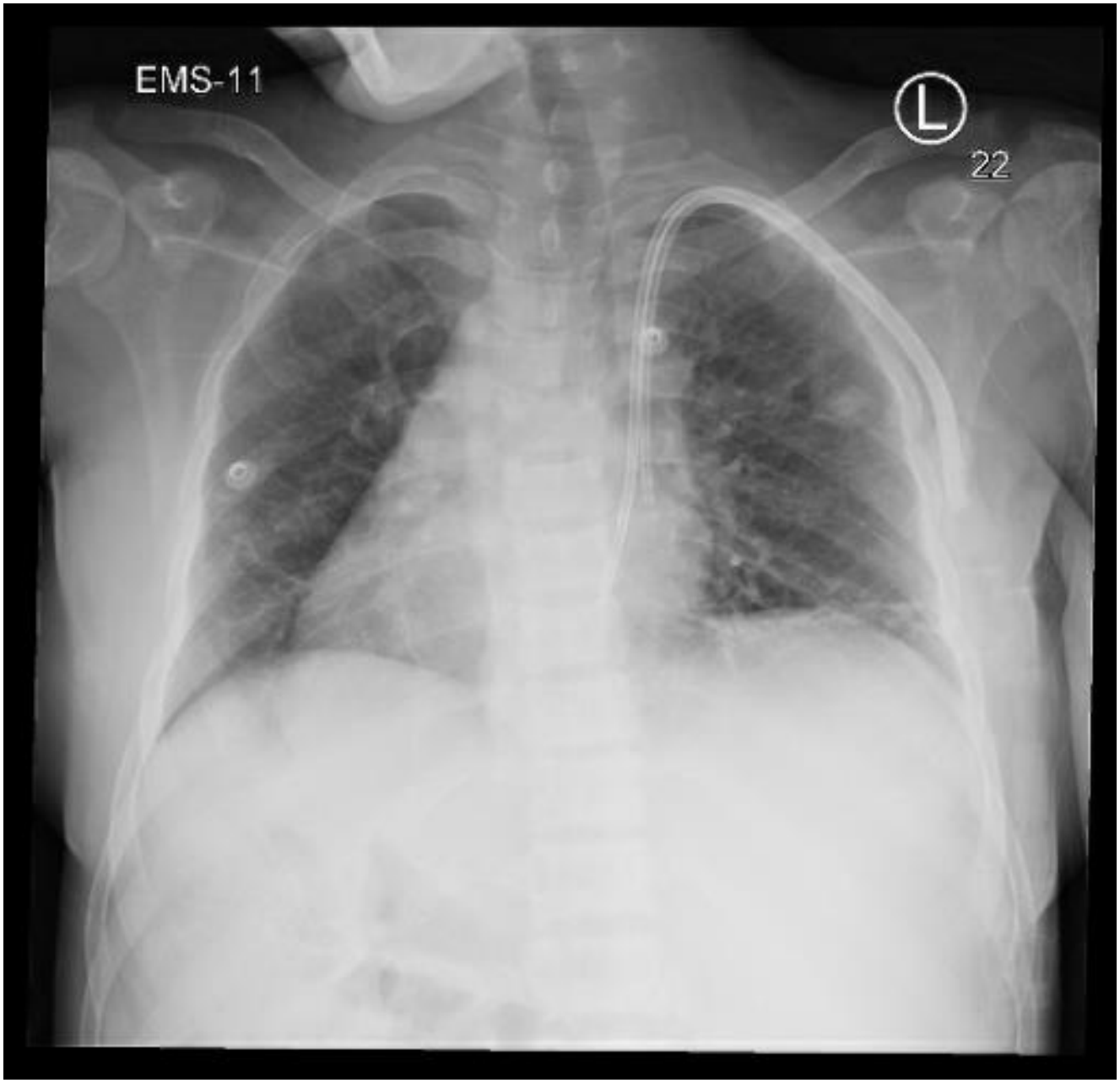


Figure 1: CXR showing dextrocardia and a left permanent catheter in situ.

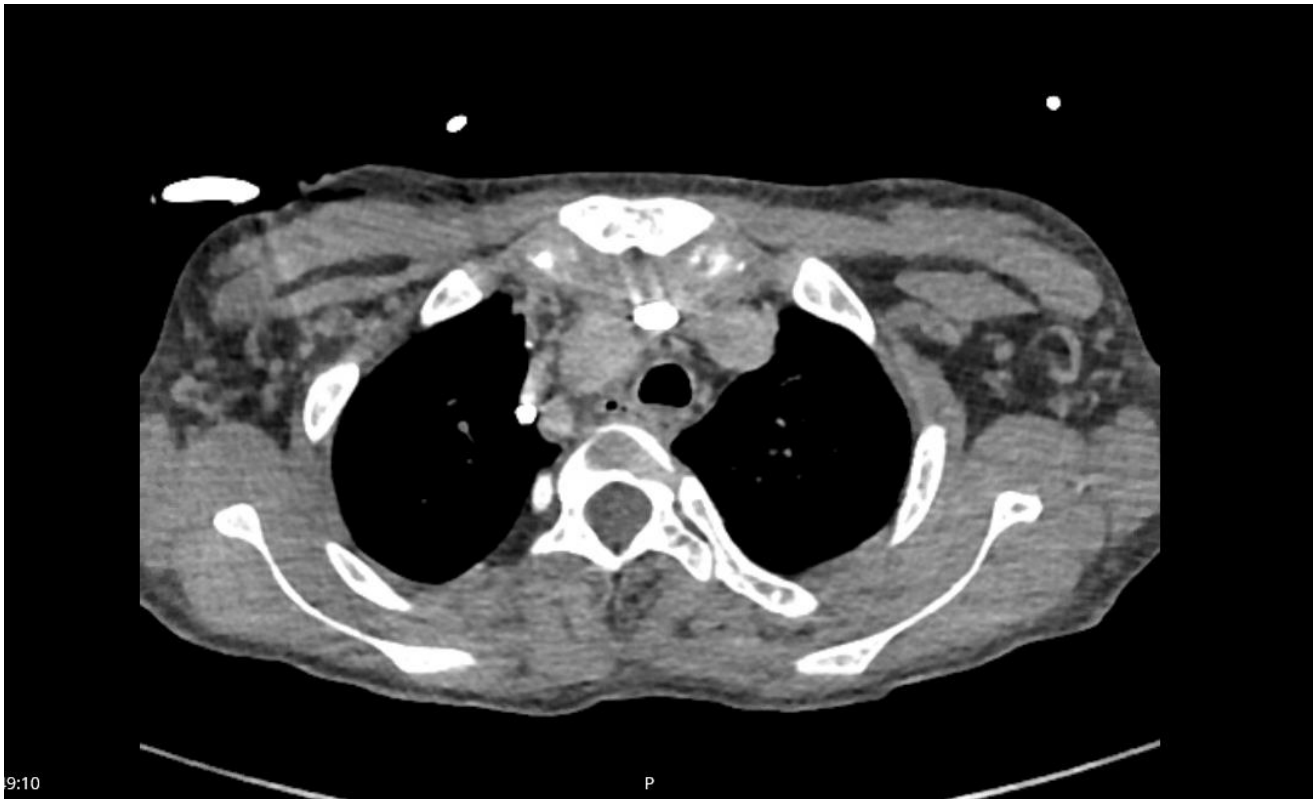


Figure 2: CT scan showing the location of the permanent catheter at IJV.