Case Report

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Intracardiac Access for Hemodialysis in Small Children: Final Options, Guidance, and Tips

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Abstract

Hemodialysis access in small children with genetic syndromes and end-stage renal failure possesses various challenges. Traditional options may quickly support an upper sternal approach over thoracotomy to enhance exposure and facilitate effective catheter placement. Overall, eight attempts were made in two cases involving both techniques. Either thoracotomy or sternotomy has inherited advantages and disadvantages. However, the sternal approach allows wider exposure, more precise catheter placement, and easier future surgeries.

Keywords: Atrium, cardiac, cardiovascular surgery, veins

Introduction

Maintaining durable and functional vascular access for children with end-stage renal disease is a challenging goal. When peritoneal dialysis is not feasible, particularly in pediatric patients, permanent tunneled venous catheters (PTVCs) are often preferred due to the complexities associated with creating and maintaining arteriovenous fistulas. However, prolonged use of PTVCs frequently leads to complications such as fibrosis, infection, thrombosis, and catheter-related fibrin sheath formation around the central vein, with small children being particularly susceptible to these issues[1-3].

Thanks to advancements in interventional techniques, unconventional vascular access options have been developed and adopted over the years[4,5]. Despite these innovations, some exceptional cases still lack viable venous access for the Seldinger technique[6]. In such situations, emergent surgical exploration remains a critical, life-saving approach.

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Transpleural or mediastinal approaches may be employed for direct cardiac catheterization. While these procedures are within the expertise of surgeons, they extend beyond simple cut-down methods and require meticulous attention.

In this article, we share our experience with eight instances of direct right atrial PTVC insertion, performed through sternotomy and thoracotomy, in two pediatric patients.

Case Presentation 1

A four-year-old female patient, weighing 16 kg, diagnosed with Emery-Dreifuss muscular dystrophy and congenital arthrogryposis multiplex, had been undergoing renal replacement therapy for over a year due to hemolytic-uremic syndrome. She was referred to our department after exhausting traditional vascular access options for PTVC placement. The patient's history included consanguineous marriage, with a heterozygous SYNE1 gene mutation (ENST00000367255.5 c.1257C>G, p.Tyr419Ter) detected. Neuromuscular development was impaired, leaving her immobile and on dual therapy for recurrent seizures. Multiple central catheter placements, port catheter placements, and hemodialysis catheter placements were attempted, but even successfully placed devices were non-durable, necessitating frequent replacements due to stenosis, thrombosis, or sepsis.

Upon initial consultation in pediatric cardiac surgery, both iliac veins showed slight re-canalization after non-surgical catheter removals, despite anticoagulant therapy. Several punctures through the jugular and subclavian veins failed because the guidewire could not advance. These challenging conditions led to the consideration of direct atrial catheter implantation. To avoid the complications associated with sternotomy, a right thoracotomy was preferred. The procedure was uneventful; the atrium was accessed via the fourth intercostal space, and the pericardium was incised. A purse-string suture with pledgets was placed on the atrial wall, and the PTVC was inserted in a manner similar to the method used in the cardiopulmonary bypass procedure (Figure 1A, 1B).

Four months post-surgery, the patient was referred due to inefficient hemodialysis. A redo surgery was performed, using a limited J-sternotomy for better exposure. The previous catheter was removed, and a new catheter was positioned to mimic the natural course of the superior vena cava, avoiding sharp angles and leaflet interference to ensure optimal suction and flow (Figure 2).

Despite these interventions, two additional surgeries were required due to sepsis and thrombosis. These subsequent replacements were successfully conducted through redo sternotomies, after which the patient achieved stable hemodialysis without further complications.

Case Presentation 2

A 4-year-old boy, weighing 14 kg, diagnosed with Denys-Drash syndrome and homozygous MTHFR C677T mutation, had been on dialysis since infancy. Over the past two years, peritoneal dialysis was discontinued due to recurrent peritonitis, sepsis, and therapy was switched to a PTVC. The patient also presented with multiple congenital anomalies, including impaired neurological development, hydrocephalus, recurrent thrombosis, cryptorchidism, and parathyrotoxicosis. All conventional vascular access sites had been exhausted, and Doppler ultrasound failed to identify any patent jugular or subclavian veins. Attempts to use femoral sites were also unsuccessful. The last inserted catheter was located in the persistent left caval vein, which was inadequate for providing sufficient hemodialysis.

Given the challenging anatomy and the previously used sternotomy approach, the surgical team opted for a thoracotomy. The initial surgical steps mirrored those of the previous case. However, to prevent catheter kinking and migration toward the tricuspid valve, a vertical, straight-line tunnel was created. This technique, while effective, introduced additional challenges: lung inspiration, could cause tension on the catheter line, necessitating a safety margin for the portion of the catheter within the atrium. Unfortunately, a miscalculation led to the distal orifice of the PTVC being positioned in the inferior vena cava, requiring an early revision for adjustment (Figure 3).

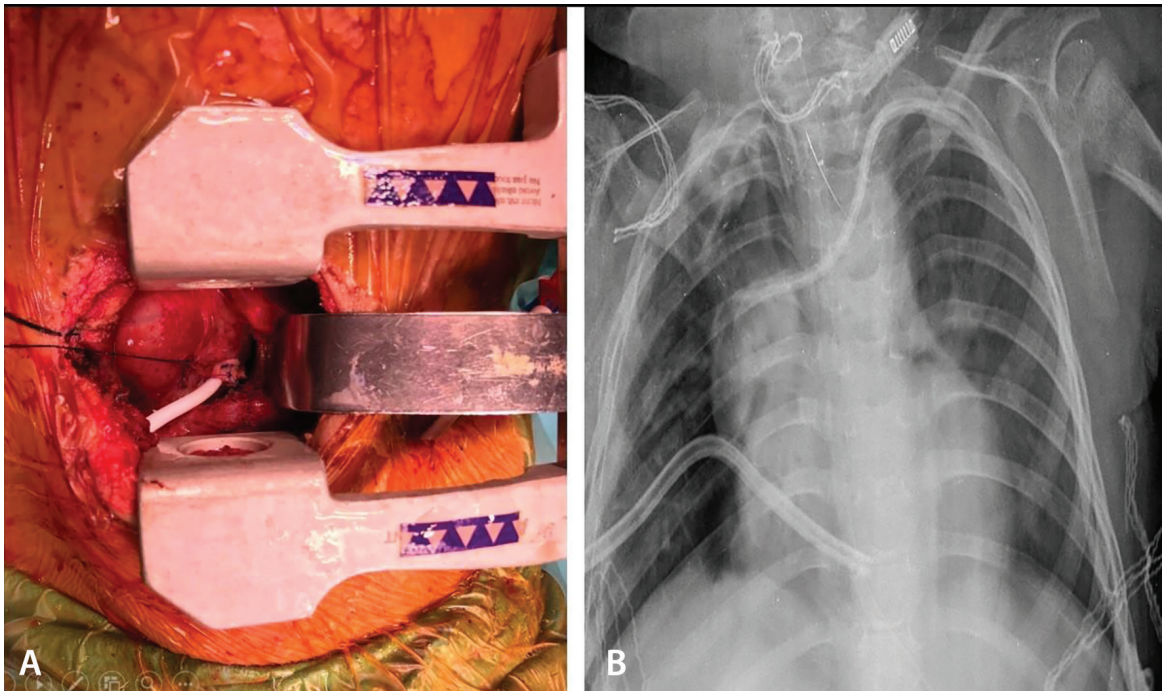

Figure 1. (A) Surgical view of the catheter directly inserted directly through right atrium, (B) X-ray view of the catheter, with the tip positioned near the tricuspid valve

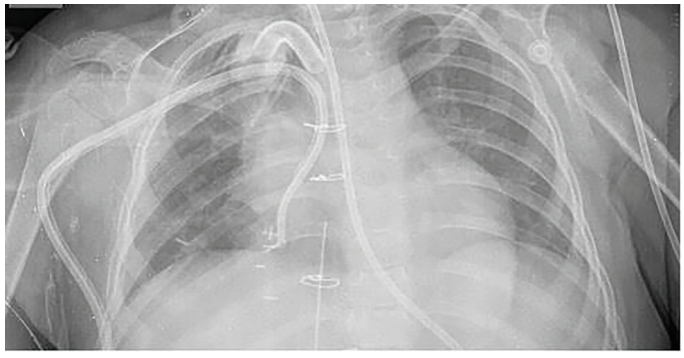

Figure 2. Right atrial catheter via thoracotomy with better course without any sharp angle

Nine months later, the patient accidentally dislodged the catheter. Fortunately, bleeding resolved spontaneously without leading to tamponade. The patient was subsequently operated on electively, and a new catheter was inserted into the right atrium via sternotomy. Due to reduced adhesion formation, the new catheter tunnel was created on the left side, following the upper portion of the clavicle, and entering the mediastinum through the suprasternal notch (Figure 4 A, B). The catheter functioned optimally thereafter.

Discussion

End-stage renal failure (ESRF) necessitates effective vascular access when transplantation or peritoneal dialysis is no longer a viable option. Although arteriovenous fistulas offer adequate blood flow with large puncture sites and reduced infection rates, their reliability in pediatric patients, especially in small toddlers, remains limited[2,3]. While most studies focus on adolescents, data on younger children, particularly those under four years of age, are scarce[7]. As a result, PTVCs are more commonly employed in pediatric populations.

The need for dialysis in children with congenital genetic syndromes and various disorders compounds the challenge, as these conditions often lead to early and rapid exhaustion of vascular access sites[8]. Given these complications, the process of selecting and managing vascular access requires a multidisciplinary approach. In many countries, pediatric nephrologists, pediatric surgeons, pediatric cardiovascular surgeons, interventional radiologists, and pediatric radiologists are

involved in managing these cases. Despite this, pediatric surgeons and interventional radiologists typically assume the primary responsibility for catheter placement globally, with the majority of published reports stemming from their expertise. Unfortunately, in Türkiye, as in many other regions, individual hospitals may develop their own protocols, which can lead to complications, particularly from a medico-legal perspective.

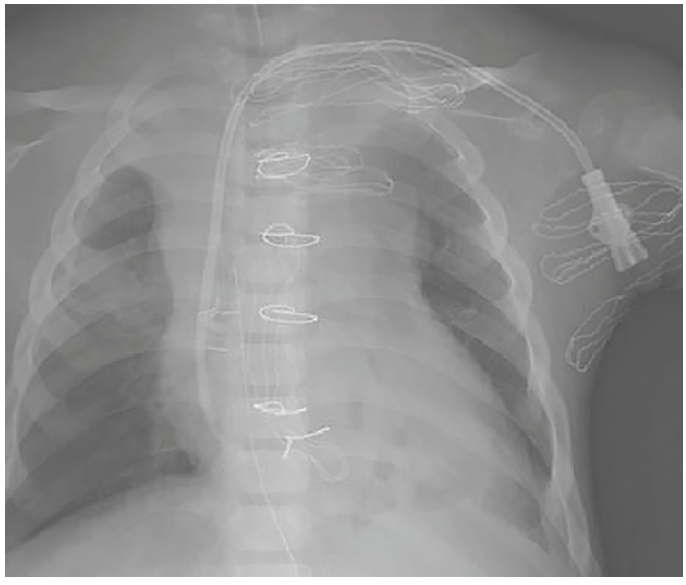

Figure 3. Revised catheter course following adjustment

We strongly advocate for the establishment of specialized, experienced catheter teams and for practitioners to exhaust all interventional options before resorting to surgery. Once all interventional methods have been exhausted, surgical intervention becomes necessary. Although non-traditional approaches such as trans-lumbar, trans-hepatic, and trans-renal catheter placements are available, only some of these approaches are feasible in young children[9,10]. Most of the existing literature on intra-cardiac PTVCs primarily reports on adult patients. Philipponnet et al.[11] reviewed 51 cases of intra-atrial catheter placement, with the youngest patient being 30 years old. This cohort was largely derived from Oguz et al.[12], which compiles and analyzes scattered reports on intra-cardiac catheterization. Notably, mortality and complication rates were significantly higher compared to those associated with interventional catheter placements or even cardiac surgery. Seven patients died within 15 days of surgery, and another six died later, a consequence likely attributed to their pre-existing poor health. Interestingly, some patients required multiple surgeries, despite reasonable follow-up periods. Only one patient from Chavanon et al.[13] underwent surgery three times.

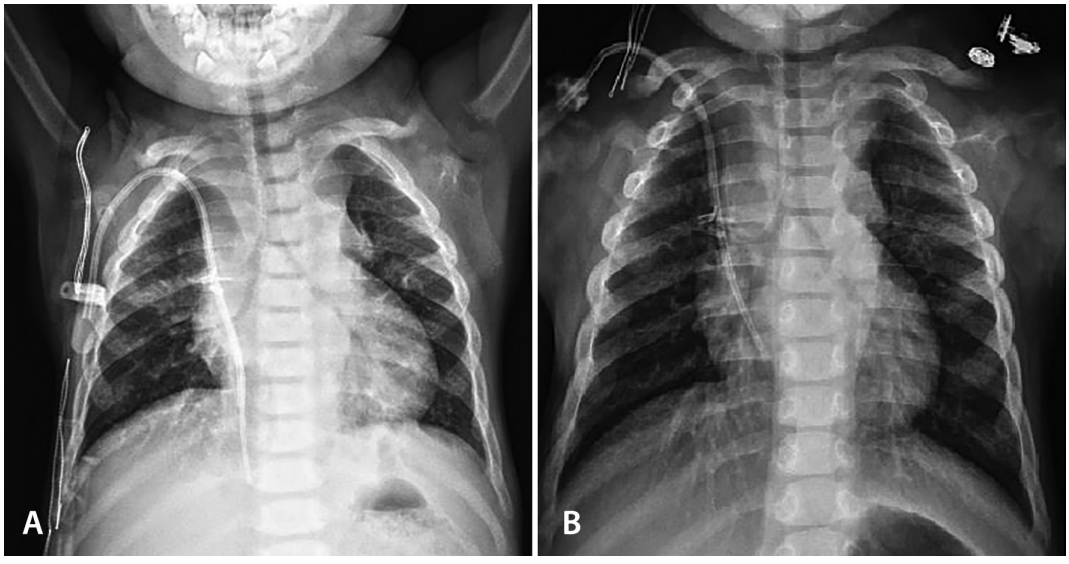

Figure 4. (A) Proper catheter alignment with no sharp angle, ensuring optimal flow. (B) The length of the catheter adjusted so as to end in the right atrium

In isolated cases of intra-cardiac catheter placement, a trans-thoracic approach is typically used[12-19], while a sternal approach is generally reserved for concomitant surgeries. In our practice, we performed four consecutive surgeries with different techniques, in both of our patients, over the past two years. Initially, we preferred thoracotomy to minimize the risks associated with sternotomy. However, this technique presents several disadvantages: pleural adhesions are more difficult to manage than those encountered during sternotomy, and the likelihood of lung injury and prolonged chest drainage is higher. Over recent decades, advancements in surgical tools and expertise have led pediatric cardiac surgeons to favor sternotomy, especially for procedures such as modified Blalock-Taussig shunts, pulmonary artery banding, and patent ductus arteriosus closure[20-22]. This increased confidence in sternotomy procedures has encouraged surgeons to reconsider sternotomy procedures for repetitive surgeries.

In addition to managing adhesions, careful attention must be paid to the safety margin required to prevent tension on the catheter during inspiration, as well as the catheter's entry angle. Improper calculations can lead to catheter migration into the caval vein or tricuspid valve, both of which impair dialysis function. Direct cardiac implantation of PTVCs generally reduces vessel-induced complications. We recommend using a catheter one size larger than usual for the child's body weight to ensure long-lasting results.

As a result, we have increasingly opted for upper sternotomy rather than thoracotomy in these cases. This approach offers better exposure, allows for more precise catheter placement, and facilitates future surgeries. Additionally, the length and trajectory of the catheter tunnel are crucial for maintaining effective hemodialysis. The closer the catheter path mirrors the natural venous flow, the better the dialysis outcomes.

We secure the catheter with sutures and pledgets at the insertion site and use silk stitches to attach the pericardium to the catheter. If possible, the catheter can be passed through the thymus to stabilize it, and in the event of accidental removal, the thymus may help in minimizing bleeding by constricting the surrounding area. However, a combination of genetic disorders, ESRF, and sepsis can impair wound healing, making it easier for even a child to dislodge the catheter with minimal effort. For this reason, the catheter must be securely fixed at the atrial entrance, pericardium, jugulum, and skin incision sites.

In summary, small children with genetic syndromes and ESRF are at increased risk for complications such as sepsis and thrombosis, which can rapidly deplete all available vascular access options. Once these interventional methods are exhausted, salvage surgery is necessary. We recommend an upper sternal approach over thoracotomy, as it provides better exposure, more precise catheter placement, and easier future surgeries. Surgeons must be mindful that these patients are often candidates for further, repetitive surgeries, and each step in the process should be carried out with this consideration in mind.

Ethics

Informed Consent: The signed informed consent forms were obtained from each parents of the patients.

Footnotes

Authorship Contributions

Surgical and Medical Practices: Gülaştı ÖF, Concept: Gülaştı ÖF, Design: Akkaya G, Data Collection and/or Processing: Gülaştı ÖF, Analysis and/or Interpretation: Akkaya G, Literature Search: Akkaya G, Writing: Akkaya G.

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