Implantable Central Venous Access Devices in Children With Metabolic Disease

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We have inserted 20 totally implantable central venous devices in 17 patients with severe metabolic disease over a 43-month span. Patient ages ranged from 2 months to 17 years (mean, 4.2 years). The underlying pathology was Gaucher's disease in six patients, vitamin D–dependent rickets type II in five, propionic acidemia in two, and methlymalonic acidemia, 3-hydroxyl-3-methylglutaryl coenzyme A (CoA) lyase deficiency, fructose 1,6 diphosphatase deficiency, and urea cycle disorder in one child each. There were seven complications (six due to catheter-related infection and one due to occlusion of the system) during a total of 7,278 patient-catheter days. The infection rate was 0.8 per 1,000 days. Six catheters were removed due to complications and two due to completion of treatment. There were no operative complications or deaths. Our experience demonstrates that a totally implantable device may be useful in children with metabolic disease who need long-term venous access. Attention should be given to minimize the infection rate to reduce the rate of catheter removal.

VENOUS ACCESS in pediatric patients has always been difficult, especially in those requiring repeated and frequent blood sampling or medication administration. Pediatric patients with metabolic disease present a special problem, since they may require frequent and urgent administration of fluids, alkalinizing agents, and drugs to prevent life-threatening complications. Long-term venous access devices (Hickman/Broviac catheters, totally implantable "ports") have become an important part in the management of patients with malignancy, fluid and electrolytes disturbances, malnutrition, infection, renal failure, hemophilia, and other chronic illnesses (eg, cystic fibrosis, sickle cell disease, etc).[1,2] We report our experience with the use of totally implantable venous access devices (TIVADs) in children with metabolic disease to assess the benefit of such a tool.

PATIENTS AND METHODS

Between February 1991 and November 1994, 20 (TIVADs) (Infuse A-Port, Infusaid, Norwood, MA) were placed in 17 pediatric patients with severe metabolic disease at the King Faisal Specialist Hospital and Research Centre (KFSH&RC), to facilitate their treatment. KFSH&RC covers a population of more than 15 million for these rare diseases. The children's medical records were reviewed with respect to age, gender, underlying disease, indication for device insertion, time from insertion, complications, outcome for the device, and reasons for removal. Before implantation of the device, a coagulation profile was obtained. All TIVADs were placed in the operating room under general anesthesia and antibiotic coverage with cefazolin (Kefazol; Lilly, England). Central venous access was obtained through cutdown on the external jugular vein in most children. A second small incision for the port implantation was made over the second intercostal space lateral to the sternum away from the breast bud. The entire system was flushed with a concentrated heparin solution (100 U/mL) before and after insertion of the catheter into the vein. C-arm fluoroscopy was used to ensure the tip of the catheter was in either the superior vena cava or the right atrium. Postoperatively, the TIVAD was immediately ready for use. All ports were flushed monthly with heparin solution, unless used for treatment.

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RESULTS

The age of the 17 children ranged from 2 months to 17 years (mean, 4.2 years). There were 11 boys and six girls.

The underlying pathology was Gaucher's disease in six patients, vitamin D–dependent rickets type II in five, fructose 1,6 diphosphatase deficiency in one, 3-hydroxy-3-methylglutaryl coenzyme A (CoA) lyase deficiency in one, methylmalonic acidemia in one, propionic acidemia in two, and a urea cycle disorder in one patient. In all cases, insertion of the TIVAD was requested by the physician managing the metabolic disease to facilitate the chronic therapy and to gain venous access in emergency situations. The six patients with Gaucher's disease required 400 U of ceredase (Alglucerase; Genzyme, Cambridge, England) every 2 weeks for life. The five patients with vitamin D–dependent rickets type II were treated with repeated calcium infusion. The two patients with fructose 1,6 diphosphatase and 3-hydroxy-3-methylglutaryl CoA lyase deficiency were susceptible to severe acidosis and hypoglycemia when given fructose-containing foods. Since this condition may elicit peripheral shock and cardiovascular arrest, immediate central venous access may be life-saving in this situation.

Patients with methylmalonic acidemia and propionic acidemia frequently developed severe acidosis requiring bicarbonate or trihydroxymethylaminomethane (THAM) and glucose administration for many days. The patient with a urea cycle disorder was vulnerable to severe hyperammonia and required intravenous arginine or intravenous Ucephan (McGaw, USA) emergently.

All procedures were performed under general anesthesia with operating times ranging from 15 to 75 minutes (mean, 59 minutes). In all patients except one, the catheter insertion was made through venous cutdown. The right external jugular vein was cannulated in 13 children, the external jugular vein in five, the left subclavian vein in one, and the right saphenous vein in one. The location of the catheter tip was in the right atrium in 14 patients, in the superior vena cava in four, in the innominate vein in one, and in the inferior vena cava in one. Thirteen Microport (8-French) and seven Button port (5-French) TIVADs were used in this group of patients. All were afebrile at the time of the surgery, except one who had a pneumonia. Their WBC counts ranged from 3 to 38 $\times 10^3/\mu L$ (mean, 13 $\times 10^3/\mu L$) and their platelet count ranged from 87 to 745 $\times 10^3/\mu L$ (mean, 367.5 $\times 10^3/\mu L$) at the time of insertion.

The total duration of implantation ranged from 10 to 1,050

days (mean, 476), with a total of 7,278 patient-days. Seven children (41%) developed complications, which necessitated removal of the implant in all except one. Four patients had septicemia, one had both pouch and systemic infection, and one had a pouch infection. Isolated organisms responsible for catheter-associated infections were *Staphylococcus aureus* in two patients, *Enterobacter agglomerus* in two, *Pseudomonas aeruginosa* in one, and *Candida albicans* in one. The catheter-associated infection rate for primary insertions was six of 17, for a frequency of 0.8 infections per 1,000 days. In all of the patients except one, the catheter infection occurred more than 100 days after insertion of the device. There was no relationship between the type of infusate and the presence of septicemia.

Only one patient had an occlusion of the system (Table 1). Ten children continue to use their TIVAD; three had their ports removed because of their infrequent use. Due to their response to oral therapy, to avoid infection, three had their device reinserted after catheter infections. Two patients died of their primary disease. There were no operative complications or deaths.

DISCUSSION

There have been multiple attempts to improve vascular access in patients who require frequent blood sampling and parenteral therapy. TIVADs are considered to be the latest refinement in this approach. TIVADs have been used in various conditions that require long-term central venous access, including malignant infections and hemophilia.[2-4]

Pediatric patients with severe metabolic disease present a challenging problem. Some need venous access for short- or long-term therapy, while others require emergent access to treat severe acidosis and shock. Physicians who care for these patients frequently request implantation of central venous devices, and we report our experience with 17 patients.

TIVADs present significant advantages over other systems like the Hickman/Broviac catheters. The totally implantable device obviates the need for dressing changes and allows vigorous physical activity, including swimming. The TIVADs were well accepted and tolerated by the children.

Various complications of TIVADs have been reported, including catheter-related sepsis, catheter leakage, occlusion, extravasation, and others.[1,5,6] Sepsis is the most serious complication and is of particular concern in our patients with metabolic diseases. The TIVAD has been shown to have an incidence of septic complications similar to or lower than that of other systems like the Hickman and Broviac catheters.[2,7-9] Six catheter-associated infections occurred in 20 catheters, for a rate of 30%. Overall infection rates in pediatric series have ranged from 10% to 60%.[6,9,10]

We encountered six infections in 7,278 days of catheter use for a rate of 0.8 per 1,000 days. This rate of infection is comparable to the experience of others.[2,4,10] The infection rate per 100 days of catheter use has been ranged from zero to five per 1,000 days of catheters use.[2,4,9,10] Considerable controversy remains concerning the treatment of catheter infection.[11] Some investigators advocate routine removal of the catheters followed by intravenous antibiotics, while others report success with antibiotic treatment through the catheter without removal.[1,7,11] The response of an infected implanted device to antibiotics therapy is variable. Unfortunately, our success rate in treating catheter-related infections with the use of parenteral antibiotics was low. Five of six infected implanted devices had to be removed due to septic complications. Pegelow et al[4] had a similar experience with infected TIVADs. They blamed their low success rate on the fact that TIVADs contain a significant dead-space volume not readily exchanged during flushing with antibiotics. In a prospective study, the device type and age of the patient were shown to have a significant affect on the rate of device-related infection, whereas the underlying disease had little effect on the infection rate.[12]

Our experience demonstrates that TIVADs may be useful in children with metabolic diseases who require long-term or urgent venous access, yet are not at risk for catheter infection and subsequent device removal.

Table 1. Patient Characteristics

Patient No.	Age at Insertion	Diagnosis	Duration of Implants (d)	Complications	Outcome
1	3 yr	Gaucher's disease	420	Systemic infection	Removed/reinserted/still in place
2	4 yr	"	630	Pocket/systemic	Removed/reinserted/still in place
3	1 yr	"	180	—	Still in place
4	7 yr	"	270	—	Still in place
5	4 yr	"	750	—	Still in place
6	1 yr	"	420	—	Still in place
7	2 yr	Vitamin D–dependent rickets type II	238	Systemic infection	Removed
8	11 yr	"	1,050	—	Still in place
9	17 yr	"	480	—	Still in place
10	1 yr 7 mo	"	450	Systemic infection	Still in place
11	13 yr	"	120	—	Still in place
12	6 yr	Methylmalonic acidemia	1,020	Occlusion	Removed
13	5 mo	Propionic acidemia	50	Pocket infection	Removed/reinserted/died
14	11 mo	3-hydroxyl-3-methylglutoryl CoA lyase deficiency	570	—	Removed*
15	2 yr	Fructose 1,6 diphosphatase deficiency	360	—	Removed*
16	5 mo	Propionic acidemia	150	Systemic infection	Removed/died
17	2 mo	Urea cycle disorder	120	—	Removed*

*Removed because of infrequent use to avoid infection.

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