



an amount of fluids was explained by a persistent intraoperative hypotension despite the absence of massive bleeding

The patient was extubated 2 hours after the end of transplantation because she was normothermic, and there was no abnormality in complete blood cell count and blood chemistry tests including blood gases. Eighteen hours after surgery, the patient presented with sudden cardiopulmonary arrest associated with airway obstruction. She had a large facial and neck edema that did not allow visualization of the larynx with a laryngoscope (Figure 2). Reintubation was rendered possible through the oral route with the use a pediatric fiberoptic bronchoscope using the guided technique [4]. She recovered after active resuscitation and mechanical ventilation. Total serum trypsinase measured in blood sample taken during resuscitation was normal. The diagnosis of superior vena cava syndrome (SVCS) was made. The SVCS was treated with drastic reduction of intravenous perfusion, changing of bedposition with head elevation, diuretics, and intravenous heparin. The patient recovered a normal consciousness on postoperative day 7. The patient's neck and facial swelling improved slowly within three weeks. In order to avoid recurrence of superior vena syndrome with airway obstruction, a preventive tracheotomy was performed on postoperative day 14. The upper airway obstruction improved sufficiently for decannulation and closure of the tracheotomy on postoperative day 28.



Figure 2 Clinical aspect of the acute superior vena cava syndrome (SVCS) with a large facial and neck edema. Reintubation was rendered possible through the oral route with the use a pediatric fiberoptic bronchoscope using the guided technique.

The immunosuppression protocol consisted of anti-thymocyteglobuline (Thymoglobuline®) induction, tacrolimus (target tacrolimus trough blood levels were between 15 and 20 ng/mL) and steroids for maintenance therapy. She was discharged at 51 days after transplantation. Twenty nine months after transplantation, the patient is well on tacrolimus (range 5 to 8 ng/mL) and steroids (prednisone: 5 mg/day), with a normal oral diet, and no episodes of rejection or enteritis (Figure 3).

Patients with intestinal failure are often managed with total parenteral nutrition (TPN) via a central venous catheter placed through one of the major veins of the neck (eg jugular, sub-clavian veins). These patients often have a history of repeated infections and thrombotic episodes secondary to these central venous accesses for TPN administration. Catheter-related thrombosis is caused by several factors including intraluminal obstruction (eg thrombus, stenosis, or

indwelling foreign body) such as in our patient, or external compression (eg tumor) [5,6]. SVC thrombosis is complicated by SVCS when the ability of collateral blood vessels to compensate for the SVC obstruction is exceeded [7].

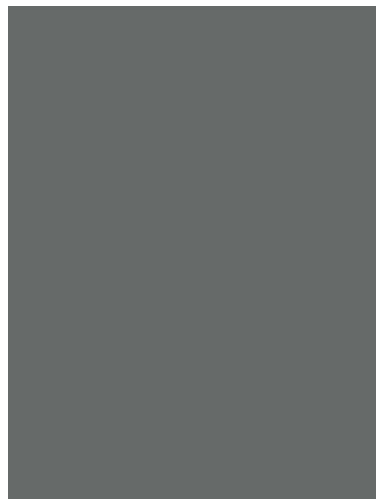


Figure 3 Mrs M. twenty nine months after transplantation. She is eating a normal oral diet.

The clinical diagnosis rate of thrombosis of the major veins of the neck associated with TPN has been reported to be 10% to 20% [8]. However, if phlebography is used, the incidence raises to 40% to 60% [9]. This makes venous thrombosis the most frequent complication of TPN [8].

Thus, in patients referred for SBT who have been maintained on long-term TPN, the presence of SVC and/or IVC obstruction must be looked for in the pre-transplant work-up even in the absence of the clinical signs of SVC syndrome. The diagnosis of chronic SVCS may often be made based on history and physical examination [10-13].

However, imaging is the mainstay for definitive diagnosis and characterization of chronic SVCS. Duplex ultrasound is the initial imaging test of choice as the technique is cost effective, noninvasive, does not involve ionizing radiation, and has high sensitivity for peripheral vein thrombosis [13]. Contrast venography may be necessary to confirm a diagnosis of SVCS if clinical suspicion remains high despite negative duplex ultrasound evaluation [5]. Magnetic resonance angiography (MRA) and computed tomography angiography (CTA) provide accurate, noninvasive methods for detecting central venous stenosis or obstruction. Both correlate extremely well with venography and can provide more detailed data regarding the level and extent of obstruction, as well as the formation of collateral pathways [6].

According to the International Intestinal Transplant Registry, 2500 small bowel transplantations have been performed worldwide [14]. However, there are very few reports of cases of intestinal transplantation performed in patients with SVC [8,15,16]. In most patients's referred for intestinal transplantation with SVC thrombosis, unconventional central venous access was used to restore venous access before transplantation [15]. Even when catheter placement was

possible, the teams underlined that in the event of severe cardiovascular instability or massive hemorrhage in the intraoperative or postoperative period, there was every chance that these catheters could have proven inadequate, even resulting in serious morbidity or mortality.

Several options have been proposed to treat *SVC* thrombosis due to benign obstructions: anticoagulation, radiation, fibrinolysis, chemotherapy, angioplasty, stenting and the more invasive surgical intraoperative venovenous bypass [17,18]. In our patient, the radiologic attempt to re-establish superior vena caval patency using a