Coronary Sinus Perforation from Placement of a LeVeen Shunt in a Child

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An unusual complication involving the LeVeen shunt is described. The patient, a two-year-old child, had chronic intractable ascites due to cirrhosis, secondary to alpha1 antitrypsin deficiency. The insertion of a LeVeen shunt resulted in perforation of the coronary sinus followed by cardiac tamponade and death.

Since its introduction in 1974, the LeVeen shunt has gained increasing acceptance as a technique for the symptomatic relief of chronic ascites refractory to medical therapy. Its utilization, however, carries significant morbidity.† We report a technical complication, not previously described, in which placement of a LeVeen shunt for the relief of ascites in an infant resulted in perforation of the coronary sinus and fatal pericardial tamponade.

Case Report

This was the sixth admission for a two-year-old white boy with documented alpha1 antitrypsin deficiency (phenotype PiZZ) who presented three weeks prior to death with vomiting, fever and tachypnea of recent onset. A diagnosis of cirrhosis was made at age one year by a liver biopsy. The patient subsequently developed sequelae of cirrhosis with portal hypertension, ascites and intermittent esophageal variceal bleeding. The ascites was initially controlled with diuretic therapy.

On this admission, he was jaundiced, irritable and in mild respiratory distress. Blood pressure was 100/62 mm Hg and temperature was 38.6°C rectally. There was hepatosplenomegaly and gross ascites. Pertinent laboratory studies showed a serum sodium level of 125 mEq/L; potassium, 4.9; chloride, 95; CO2, 19; BUN, 14; and albumin, 2 g/L.

Chest x-ray examination showed a large right pleural effusion and a slightly enlarged cardiac silhouette.

A diagnosis of fluid overload and sepsis was made and efforts directed at controlling these. Two weeks later, the patient developed oliguria, and was given intravenous albumin and furosemide, in addition to the spironolactone and triamterene he had been receiving regularly. Despite this regimen, the patient remained oliguric, with continuing weight gain, tense ascites and increasing peripheral edema, hyponatremia, hypokalemia and azotemia. Immersion therapy failed to improve absorption of ascites or promote diuresis. In view of his pre-renat azotemia and poor prognosis, it was elected to place a peritoneovenous shunt. Nineteen days after admission, the patient underwent insertion of a LeVeen shunt under local anesthesia. The shunt was placed on the right side, with the valve underneath the

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Figure 1. Posterior surface of the heart with the shunt tube in place in the coronary sinus. A small hemorrhage with clotted blood at the tip of the sinus can be seen at the left rectus muscle, and the venous limb was secured in the right internal jugular vein. During insertion of the shunt, the neck was hyperextended and rotated to the left until positioning of the venous limb was completed; the venous limb was shortened as to reside appropriately in the area of the junction of the superior vena cava and right atrium. The immediate postoperative period was unremarkable. However, the following day, less than 24 hours after surgery, the patient cried out in bed and became unresponsive. Efforts at resuscitation were unsuccessful.

At autopsy, there was a massive serous ascites of 1,970 ml. The liver showed macronodular cirrhosis and the spleen, congestive splenomegaly. The findings in the heart were of most interest. The pericardial sac was tense, with 90 ml of fresh blood within it. A small, irregular tear 2 to 3 mm in length with surrounding soft tissue hemorrhage was seen in the epicardium in the atrioventricular groove on the posterior surface of the heart (Fig 1 and 2). When the

Figure 2. Schematic representation of the findings in the heart. A. Right lateral view of the heart externally showing the swelling and tear of the coronary sinus from the venous end of the shunt tube. B. A cut view of the right atrium showing the course of the shunt tube from the superior vena cava into the coronary sinus. CS = coronary sinus; CSO = coronary sinus ostium; IVC = inferior vena cava; RA = right atrium; SVC = superior vena cava.
heart was opened, the area of the epicardial tear and hemorrhage was traced to the coronary sinus, about 1 cm from its entry into the right atrium. A substantial length of the distal portion of the shunt tubing was lying freely within the right atrial cavity. While it could be easily passed into the coronary sinus ostium, to abut against the tear, it could not be passed into the sinus beyond this point. Microscopic findings confirmed the presence of hemorrhage into the tissues at the site of the tear. It was surmised that the patient's death had been due to acute pericardial hemorrhage and tamponade from perforation of the coronary sinus by the venous limb of the LeVeen shunt.

Discussion

As use of peritoneovenous shunting for intractable ascites has gained increasing acceptance, reports of complications have, predictably, become more frequent. Greig et al had reported a complication rate of 75 percent in their series, somewhat higher than the experience of LeVeen and colleagues. The majority of complications include fever, wound infection, septicemia and peritonitis, asymptomatic coagulopathies or disseminated intravascular coagulation, congestive heart failure or arrhythmias, variceal hemorrhage, electrolyte disturbances, liver function deterioration or hepatic coma, and small bowel obstruction. These complications and their rates of occurrence are understandable in light of the severe nutritional and immunologic impairment, fluid-electrolyte imbalance and marginal hepatic reserve of the patients undergoing peritoneovenous shunting. The mechanisms of these complications are often elusive, particularly regarding the coagulopathies, and require further elucidation if use of the LeVeen shunt is to gain broader acceptance.

A number of complications, on the other hand, are related to technical difficulties, which should be more readily surmounted. Generally, these are related either to malposition or occlusion of the shunting mechanism, or to ascites leak. LeVeen has reported sporadic instances of pneumothorax or transient damage of the recurrent laryngeal nerve during replacement. Thrombosis of the superior vena cava has also been described.

Proper placement of the shunt mechanism, particularly the venous limb, is crucial to its function. Positioning of the venous limb of the shunt within the internal jugular vein obviates kinking and occlusion of the shunt. The ligature around the distal aspect of the internal jugular vein obviously should not occlude the Silastic tubing of the venous limb. Yet if not tied securely, it can result in migration and dislodgement of the shunt out of the vena cava, postulated to be due to traction on the tubing by intestinal or omental motility. Holcroft et al described a case in which the venous limb was advanced too far, placing the distal lumen in the infra-diaphragmatic vena cava, with resultant loss of the pressure gradient essential to shunt function. Occlusion of the shunt by fibrin or peritoneal cells is uncommon and requires replacement of the valve.

Experience with the use of the shunt in children is limited. The vast majority of reported cases have involved adults. LeVeen alluded to the placement of the shunt in the management of chylous ascites in two children without untoward effect. In our case, the LeVeen shunt was placed with the hope of improving ascites refractory to medical therapy, and reversing the pre-renal component of this azotemia. During placement of the shunt, the patient's neck was hyperextended to gain access to the right internal jugular vein, which was easily mobilized and of a sufficient caliber. The venous limb of the shunt was trimmed after careful measurement of the distance corresponding to the right atrium and inserted into the vein without difficulty; the limb was secured in place with two ligatures placed distally. No x-ray film contrast study for shunt placement was done, as the shunt was functioning well. The following morning, the patient sustained a cardiopulmonary arrest which was irreversible. At autopsy, there was hemopericardium, from perforation of the coronary sinus near its ostium. The proximity and diameter of the shunt tubing coincided with the perforation, implicating the venous limb of the shunt as its cause. It is postulated that in hyperextending the neck of the child to gain access to the jugular vein, misjudgment as to the length of the tubing required to enter the right atrium ensued. In placing the neck out of hyperextension, the trimmed end of the venous limb probably was passed more distally. Since the confluence of the superior vena cava, right atrium and ostium of the coronary sinus are in very close proximity in a child, distal passage of the shunt evidently caused the tubing to enter the coronary sinus, later resulting in perforation, hemopericardium and death.

This unfortunate occurrence has not been described in previous reports on the complications of the LeVeen shunt. We believe that the shunt is efficacious in the treatment of ascites in selected patients refractory to medical therapy in both pediatric and adult patients. For these reasons, we have reported this case to alert others to the potential for this complication. We also concur with Holcroft et al and Vaida and Lauciuzs who advocate the use of a radiopaque marker on the Silastic tubing to aid in placement and subsequent identification of the shunt as an adjunct to careful clinical monitoring. Greig et al routinely perform shuntograms at the time of shunt insertion to confirm correct placement. We feel that it is imperative to verify shunt position at the time of placement, particularly in pediatric patients and those with small body habits, if shunt function is to be assured and complications of the enormity described in our patient are to be avoided.

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References

Endobronchial Necrobiotic Nodule Antedating Rheumatoid Arthritis*

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We describe the previously unreported occurrence of an endobronchial necrobiotic nodule. This obstructing lesion of the left mainstem bronchus mimicked bronchogenic carcinoma and antedated clinically apparent seronegative rheumatoid arthritis by seven years.

At least five different types of lung disease can occur in association with systemic rheumatoid disease. These include: 1) fibrosing alveolitis; 2) necrobiotic nodules; 3) pleuritis and pleural effusion; 4) obliterator bronchiolitis; and 5) rheumatoid vasculitis.1 Although the diagnosis of rheumatoid lung disease may be a simple matter when clinically apparent rheumatoid arthritis is present, pulmonary manifestations may antedate the occurrence of symptomatic arthritis and thus present a diagnostic dilemma.2 We describe in this report the unusual occurrence of a necrobiotic rheumatoid nodule within the left mainstem bronchus seven years prior to the onset of clinical rheumatoid arthritis. This nodule mimicked bronchogenic carcinoma and was accompanied by concentric diffuse bronchostenosis.

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Figure 1. Low power photomicrograph of endobronchial mass. Note chronic inflammation, fibrosis, and pseudogranuloma formation.

CASE REPORT

A 48-year-old white woman presented in June, 1970 with wheezing following an upper respiratory infection. She denied dyspnea, cough, hemoptysis, or weight loss, but had a ten pack-year smoking history. On physical examination, she had grossly audible wheezing which on auscultation was localized to the left anterior chest. A chest roentgenogram revealed fullness of the left hilum. Tomography of the bronchial tree demonstrated narrowing of the left mainstem and upper lobe bronchi. The clinical impression was bronchogenic carcinoma. Rigid bronchoscopy demonstrated a friable exophytic mass partially occluding the left mainstem bronchus. The remainder of this bronchus and the upper lobe bronchi were diffusely narrowed. Biopsies were interpreted as showing chronic granulomatous inflammation and fibrosis (Fig 1). Routine and special stains of the specimen revealed no pathogens and cultures were negative.

In June, 1977 the patient experienced the onset of painful swelling and stiffness of her hands bilaterally as well as swelling and tenderness of the feet. The stiffness was worse in the morning, lasted for a few hours, would tend to recur towards late afternoon and was accompanied by generalized fatigue. She denied dry eyes, dry mouth, fevers, skin eruption, gastrointestinal symptoms or back pain. Physical examination revealed bilateral swelling, warmth, and synovial thickening of the wrists, the first and second MCP joints, and the first three PIP joints. There was swelling, warmth

Figure 2. High power view of mass. Note necrobiosis of collagen, palisaded cells, and chronic inflammation typical of a necrobiotic nodule.