esophagograms (Fig 2).

The patient was gradually able to tolerate solid foods. Three weeks after the last operation, he was discharged to his home. In the next few weeks the patient gained 9.1 kg (20 lb) in weight; however, while at home, he suffered a fatal coronary attack. Examination of the esophagus at autopsy revealed an intact esophageal lumen without stricture formation at the site of initial rupture.

DISCUSSION

It is generally agreed that the treatment of choice in esophageal perforation, when detected within six to eight hours, is closure by primary suture, as stressed by Samson. In the intermediate phase of perforation, extending up to about 24 hours, several procedures have been suggested, including the "onlay gastric" patch procedure of Thal and Hatafuku, either alone or in combination with fundoplication.

Yet there are conflicting opinions regarding the treatment of esophageal rupture complicated by fistulous tracts, empyema and other pulmonary problems, sepsis, and marked malnutrition. In these patients, death is almost inevitable unless the infection can be controlled and some form of alimentation restored. For the correction of such problems, Johnson and associates utilized the exclusion technique as applied to congenital tracheoesophageal fistula. The initial procedure consisted of a completely diverting cervical esophagostomy with closure by suture of the thoracic esophagus, proximally and distally to the perforation. Later, continuity was restored, usually by interposition of stomach or colon. Recently, Urschel et al published a report of six cases, applying the principle of exclusion and diversion in continuity, with preservation of the esophagus and reconstruction at a later date.

Salient features of the method used in our patient were as follows: (1) excision of the empyema sac with decortication and adequate drainage; (2) exclusion of the distal thoracic esophagus by ligation without closure or disturbance of the perforation; (3) diversion of secretions by cervical esophagostomy in continuity with Foley catheter placed intraluminally; and (4) feeding gastrostomy or jejunostomy for alimentation. Disappointment with the fact that the esophagus is not preserved in some methods used to treat delayed esophageal rupture led us to adopt the plan detailed herein. We believe that the principle of placing injured tissues at rest applies equally as well to the treatment of esophageal perforation. The complete diversion of secretions away from the injured site is in keeping with this principle.

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Situs Inversus, Subaortic and Subpulmonic Stenosis, Ventricular Septal Defect, and Single Coronary Artery*

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The unusual occurrence of situs inversus totalis, ventricular septal defect, hypertrophic subaortic and subpulmonic stenosis, and single coronary artery in a 38-year-old man is presented. The clinical course was remarkably mild, as documented by data from 23 years of study including four cardiac catheterizations. At age 35 years, however, syncope, chest pain, and marked elevation of right ventricular pressure prompted complete surgical repair of the left and right ventricular outflow tract obstructions and closure of the septal defect. Three years after surgery the patient continues to lead an active life without symptoms. The unusually mild course can be attributed to the natural barding effects of the subpulmonic stenosis which prevented irreversible pulmonary hypertension.

Hypertrophic subaortic stenosis associated with situs inversus was first reported in 1965. Two additional case reports were described in 1975, one of whom also had subpulmonic stenosis. We present a 38-year-old man with situs inversus totalis associated with subaortic and subpulmonic stenosis, ventricular septal defect, and single coronary artery, followed-up by us for 23 years. This unusual combination of cardiac anomalies, to our knowledge, is unique.

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CASE REPORT

A 15-year-old Puerto Rican boy was first seen by the cardiology service of Bellevue Hospital in 1952 for evaluation of congenital heart disease. There was a past history of complete situs inversus and a holosystolic murmur noted since age six. Cyanosis was never present and he had good exercise tolerance and normal development. Family history was unremarkable.

Examination revealed a normal appearing youth with a pulse of 98 beats per minute and blood pressure of 96/64 mm Hg. The heart was right-sided and a grade 4/6 holosystolic murmur and thrill were noted at the lower right sternal border. The initial cardiac diagnosis was a cyanotic tetralogy of Fallot. Situs inversus totalis was confirmed by upper gastrointestinal series, chest x-ray film and ECG (Fig 1). Arterial oxygen saturation was normal. Right heart catheterization using the left antecubital vein was performed for the first time in 1952 (Table 1), and showed an interventricular septal defect and infundibular stenosis with a 20 mm Hg gradient across the pulmonic outflow tract. Pulmonary artery, right atrium, right ventricle, and great vessels were mirror image normal by angiography.

A second cardiac catheterization using the Judkins technique was performed at age 30 because of precordial pain and exertional dyspnea. Results were similar to those of the first study (Table 1). Hydrogen arrival times and oxygen saturations confirmed a high VSD with left-to-right shunt.

Table 1—Cardiac Catheterization Data

<table>
<thead>
<tr>
<th>Pressures (mm Hg)</th>
<th>1952</th>
<th>1966</th>
<th>1/24/72</th>
<th>5/22/72</th>
</tr>
</thead>
<tbody>
<tr>
<td>Right ventricle</td>
<td>74/5</td>
<td>73/7</td>
<td>95/8</td>
<td>31/5</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>59/36</td>
<td>44/19</td>
<td>37/17</td>
<td>23/8</td>
</tr>
<tr>
<td>Pulmonary wedge</td>
<td>—</td>
<td>6</td>
<td>14</td>
<td>—</td>
</tr>
<tr>
<td>Left ventricle</td>
<td>—</td>
<td>100/5</td>
<td>170/5</td>
<td>—</td>
</tr>
<tr>
<td>Aorta</td>
<td>125/73</td>
<td>107/73</td>
<td>170/75</td>
<td>—</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Right atrium</th>
<th>A Wave</th>
<th>—</th>
<th>5</th>
<th>7</th>
</tr>
</thead>
<tbody>
<tr>
<td>V Wave</td>
<td>—</td>
<td>2</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>Mean</td>
<td>5</td>
<td>1</td>
<td>3</td>
<td></td>
</tr>
</tbody>
</table>

Systemic blood flow (L/min) 3.9 — 3.2 3.4
Pulmonary blood flow 9.3 — 8.3 3.4

At age 35, he developed pressing chest pain, exertional dyspnea, and syncope, and was restudied by a third cardiac catheterization (Table 1) using the Judkins technique. The pressure gradient across the pulmonary outflow tract was now 50 mm Hg and right ventricular pressure was now equal to left ventricular pressure of 100 mm Hg. Aortic root angiogram revealed a normal ascending and descending aorta without aortic regurgitation. The aortic valve was normal. A single main left coronary artery giving rise to three large coronary arteries representing the equivalent of the LAD, circumflex and RCA was observed.

In view of the increased right ventricular pressure and his worsening clinical state, open heart surgery was performed. Mirror image dextrocardia and single left coronary artery were confirmed, and marked biventricular hypertrophy was found. Right ventriculotomy revealed hypertrophic, muscular, infundibular stenosis and a large moderator band. On the left, hypertrophic subaortic stenosis was present. Both hypertrophic outflow tracts were resected. Microscopic examination of the tissue revealed hypertrophic cardiac muscle. A high interventricular septal defect of 2 cm in diameter was closed by a Dacron patch. The postoperative course was uneventful.

Three weeks after surgery a murmur of aortic regurgitation was heard for the first time. Recatheterization showed

Figure 1. ECG showing negative P, QRS, and T wave in lead I characteristics of dextrocardia. The right-sided precordial leads (lowest strip) demonstrate an unusually tall R wave in V4R and negative T wave consistent with left ventricular hypertrophy. The upper and middle strips are standard ECG leads. Paper speed is 25 mm/sec. Gain, 10 mm = 1 mV.

Figure 2. Chest x-ray film showing dextrocardia, cardiomegaly, and prominent pulmonary vasculature.
marked improvement in the right-sided pressures, and a peak gradient of 8 mm Hg across the left ventricular outflow tract (Table 1). Aortic root angiogram showed moderate aortic regurgitation with prolapse of the posterior aortic cusp. Hydrogen arrival times showed absence of shunting. Chest x-ray film postoperation is shown in Figure 2. Over the last three years he has remained free of chest pain and syncope.

**Discussion**

Situs inversus totalis is a rare congenital anomaly, as demonstrated by mass radiographic screening in Norway which yielded an incidence of 1 in 10,000. Single coronary artery is also rare; a recent review suggested an incidence of 4 in 10,000. The chance of these two lesions occurring simultaneously is thus about 4 in 1 x 10^6. Hypertrophic subaortic stenosis is uncommonly associated with other cardiac anomalies. The constellation of defects in the above case is therefore unique.

In this patient, the unusually mild, longterm, clinical course is attributed to infundibular stenosis resulting in a natural “banding” of the pulmonary outflow tract. This is analogous to surgical banding of the pulmonary artery in order to prevent massive pulmonary blood flow and pulmonary hypertension in neonates deemed too young for closure of a ventricular septal defect.

The weakness, chest pain, and syncope which developed in our patient at age 35 are attributed to the combination of left ventricular outflow obstruction and ventricular septal defect rather than to any effects of the single coronary artery which was widely patent angiographically. Complete surgical correction of the defects resulted in marked improvement in the hemodynamic and clinical states.

The development of aortic regurgitation after surgery was a result of closure of the high ventricular septal defect and was not present prior to surgery.

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**Prophylactic Irradiation of the Lung in Ewing’s Sarcoma**

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A patient with Ewing’s sarcoma of the left scapula received prophylactic radiotherapy to the left lung following wide excision of the tumor. While the left lung remained free of tumor, there was metastatic invasion of the right lung within ten months of the initial diagnosis of the disease. Since Ewing’s sarcoma frequently metastasizes to the lungs, it is suggested that prophylactic irradiation of the lung may be of value in the prevention of metastases.

Ewing’s sarcoma, or diffuse endotheioma of bone, is an uncommon primary malignant neoplasm. The traditional treatment for patients with localized Ewing’s sarcoma has been either surgical or radiation therapy of the primary tumor. Controversy has existed as to the preferable method of treatment, despite the high frequency of local control with both, cure is seldom effected because of rapid dissemination resulting from early hematogenous spread. The high probability of occult metastases being present at the time of diagnosis has stimulated the search for adjuvant systemic therapy designed to eradicate microscopic foci of metastases, namely, systemic chemotherapy or total-body irradiation.

This report describes the effect of prophylactic irradiation of the lung in a patient with Ewing’s sarcoma.

**Case Report**

A 26-year-old woman was diagnosed in January 1973 as having Ewing’s sarcoma. The primary site was the left scapula, with no evidence of distant metastases. Surgery, which included wide excision of the tumor and adjacent soft tissue, was followed by radiotherapy (Telecobalt unit). During March and April 1973, the patient received 3,000 rads (posteroanterior at a depth of 4 cm) to the left hemithorax (field size, 20 x 16 cm) in three weeks, and an additional 3,000 rads (posteroanterior at a depth of 4 cm) to the left scapular region (field size, 14 x 12 cm) in three weeks. Subsequently, by the end of April 1973, chemotherapy at dosages of 1.5 mg of vincristine per square meter of body surface area and 400 mg of cyclophosphamide per square meter of body surface area was given once weekly for six weeks and then once biweekly. A chest x-ray film in July 1973 was normal (Fig 1).

In September 1973, transient signs and symptoms of radiation-induced pneumonitis were noted. Ten months after surgery (November 1973), a single small metastatic lesion appeared in the right lung, and despite further intensive chemotherapy with adriamycin (30 mg/sq m of body surface area) and vincristine (1.5 mg/sq m of body surface area), an increase in the size and number of lesions in this lung could be seen on a repeat chest x-ray film in July 1974 (Fig 2).

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