for one year. Pleural effusion in this case was due to a large plasmacytoma of the chest wall.

Gabriell reported a case of pleural plasmacytoma in a 50-year-old man with chest pain. At autopsy the pleura was replaced by a tumor measuring 1 to 1.5 cm in diameter. The tumor did not invade the lung.

Favis and co-workers described the case of a 60-year-old woman with bilateral pleural effusion and many plasma cells in the pleural fluid, probably due to pleural plasmacytoma with subpleural and pulmonary parenchymal myelomatosis.

Gabriell reported a case of a 63-year-old woman with massive pleural effusion. The cytopathologic examination of pleural fluid and aspirated lung tissue showed a prevalence of plasma cells and paper electrophoresis of pleural fluid revealed a gamma globulin characteristic of myeloma in the beta range.

Edwards and Zawadzki reported a case of pleural effusion in a 69-year-old man with multiple myeloma; involvement of the pleura was proved at postmortem examination.

Intrathoracic structures such as lung, pleura, thymus, bronchi, hila, and mediastinum are well supplied with lymphatic tissues. The plasma cells are thought to be derived from lymphocytes or blasts in the lymphoid tissue, so this may explain why the pleura may be involved in multiple myeloma.

REFERENCES
1 Kudrewetzky B: Cited by Hayes et al. Ztschr Heilk 13:300, 1982
6 Hellwig CA: Extramedullary plasma cell tumors as observed in various locations. Arch Path 36:85-111, 1943
8 Herskovic T, Andersen HA, Bayrd ED: Intrathoracic plasmacytoma; presentation of 21 cases and review of literature. Dis Chest 47:1-6, 1965

Ebstein's Malformation of the Tricuspid Valve Associated with Valvular Stenosis and Cor Triatriatum*

Arthur H. L. From, M.D.; William F. Mazzitello, M.D., F.C.C.P.; Allen S. Judd, M.D.; and Jesse E. Edwards, M.D.

The case of a 39-year-old man with Ebstein's malformation, valvular pulmonary stenosis, atrial septal defect, ostium secundum type, and cor triatriatum is reported. The patient had cyanosis throughout his life but only moderate functional impairment of the heart. The mode of death suggested paradoxical embolization with the right atrium a possible source of the emboli.

Ebstein's malformation of the tricuspid valve tends to occur as an isolated condition except for the common association of a patent foramen ovale. Nevertheless, several other conditions may be associated with Ebstein's anomaly. These include ventricular septal defect, pulmonary stenosis with or without a ventricular septal defect, pulmonary atresia with intact ventricular septum, and several transposition syndromes. Most of these anomalies are not commonly associated with Ebstein's malformation, and pulmonary valvular stenosis is rare in the absence of ventricular septal defect. The purpose of this report is to describe the clinical and pathologic features of a case of Ebstein's malformation associated with a rare combination of anomalies, namely, valvular pulmonic stenosis and cor triatriatum.

CASE REPORT

Clinical Features

The patient, a 39-year-old man, was admitted to St. Mary's Hospital for diagnostic cardiac catheterization. He had a lifelong history of mild cyanosis and a cardiac murmur. The major complaints were those of mild exertional dyspnea, increased ease of fatigueability, and questionable exertional precordial distress.

Pertinent physical findings included moderate cyanosis, clubbing of the fingers and toes, a left precordial bulge, a left parasternal lift, and a systolic ejection murmur, grade 3/4, which was heard best along the left sternal edge. The first cardiac sound was normal, and the second was thought to be single. There were no diastolic murmurs or gallop sounds. The concentration of hemoglobin was 19 gm/100 ml of blood. Urinalysis gave negative results.

A thoracic roentgenogram (Fig 1) revealed a mild degree

*From the Departments of Medicine and Pathology, The University of Minnesota Hospitals and St. Mary's Hospital, Minneapolis, Minnesota, and the Department of Pathology, Miller Division, United Hospitals, Inc., St. Paul. This study was supported by Public Health Service Research Grant 5 RO1 HL05694 and Research Training Grant 5 TO1 HL08570 from the National Heart and Lung Institute, by the Minnesota Heart Association and the Minnesota Medical Foundation.

Reprint requests: Dr. Edwards, United Hospitals, Miller Division, 125 West College Avenue, St. Paul 55102

CHEST, VOL. 64, NO. 2, AUGUST, 1973
of cardiomegaly and prominence of the pulmonary artery. The lungs appeared normal. Increased pulmonary flow was not suggested. An electrocardiogram (Fig 2) demonstrated right axis deviation and complete right bundle-branch block. P pulmonale was not present.

The results of catheterization of the right heart were as follows: The right atrial mean pressure measured 6 mm Hg. The right ventricular systolic pressure was 86 mm Hg, and a peak systolic gradient of 46 mm Hg was present between the high right ventricle and the main pulmonary artery. Right ventricular end-diastolic pressure was elevated to 12 mm Hg. Left ventricular systolic pressure was 120 mm Hg and the end-diastolic pressure was 12 mm Hg. The mean pressure in the pulmonary artery was 13 mm Hg. The oxygen saturation of blood from the main pulmonary artery was 80 percent, and that of blood from the brachial artery was 93 percent. Indicator-dilution curves demonstrated a right-to-left shunt. Injection of contrast material into the right atrium showed a right-to-left transatrial shunt and a dome-shaped pulmonary valve associated with poststenotic dilatation of the pulmonary artery. The right atrial and right ventricular chambers appeared to be within normal limits as to size, and, although the right ventricular chamber appeared unusual, the nature of the abnormality was unclear.

**Figure 2.** Electrocardiogram showing right bundle-branch block and right axis deviation. Note no evidence of P pulmonale.

**Figure 3.** Diagram summarizing pathologic features of case. SVC = superior vena cava; IVC = inferior vena cava; RA = right atrium; “RV” = atrialized portion of right ventricle; TVO = true tricuspid orifice; ATO = accessory tricuspid orifice; PV = stenotic pulmonary valve; ALA = accessory left atrial chamber; LA = true left atrium; LV = left ventricle.

After returning home on the day of discharge (one day following the catheterization), the patient suddenly fell over while sitting in a chair, lost consciousness, and remained comatose until his death two days later.

**Pathologic Features**

Pertinent necropsy findings were confined to the central nervous system, the kidneys and the heart. The lumen of the right common carotid artery was occluded by a thrombus which began beyond the siphon and extended into the right middle and anterior cerebral branches. Approximately 80 percent of the right cerebral hemisphere was infarcted. No other lesions or significant atherosclerotic vascular changes were identified in the brain.

Examination of the kidneys revealed three small hemorrhagic infarcts, the largest of which measured approximately 1 cm in diameter. The renal vasculature was grossly free of emboli or atherosclerotic changes.

The heart was moderately enlarged, weighing 400 gm. Several malformations were present including Ebstein's malformation of the tricuspid valve, pulmonary valvular stenosis, and cor triatriatum (Fig 3).

The tricuspid valve showed major downward displacement of its septal and posterior leaflets so that a significant part of the right ventricle was atrialized, and formed along with the right atrium the right-sided receiving chamber (Fig 4). The ventricular aspects of the septal and posterior leaflets were attached over their entire extent to the right ventricular wall. The anterior leaflet was large. Although its basal attachment was normal, its free edge was in part attached to the right ventricular wall by poorly developed chordae and papillary muscles. This yielded a narrow tricuspid orifice, measuring about 1.0 cm in diameter. A small accessory orifice, guarded
The deformity of the tricuspid valve was responsible for major reduction in the size of the right ventricle. The right ventricle as a functioning unit consisted of hardly more than the infundibular portion. The pulmonary valve had only one commissure and exhibited a dome-shaped deformity. The valvular orifice measured 0.6 cm in diameter (Fig 5). An atrial septal defect at the fossa ovalis measured approximately 1.5 cm in diameter (Fig 4). Inspection of the interior of the left side of the heart revealed that the left atrium was divided into two chambers, an upper chamber to which the four pulmonary veins connected and a lower chamber which communicated with the mitral valvular orifice, the left atrial appendage and the atrial septal defect. A diaphragm separated the two left atrial cavities. An opening in the diaphragm, which was approximately 1.5 cm in diameter, allowed communication between the two chambers of the left atrium (Fig 6). The left ventricle, mitral valve, and great vessels were normal, and no ventricular septal defect was noted. A small quantity of friable thrombotic material was loosely attached to the wall of the right atrium.

Microscopic examination confirmed the presence of renal and cerebral infarcts.

**Comment**

The combination of anomalies just described has, to the best of our knowledge, not been previously reported and, although complex, did not cause more than moderate symptoms even though the patient was 39 years of age. This was apparently due to the fact that the lesions caused only moderate obstruction to flow into the pulmonary artery, with no impediment to pulmonary venous drainage. Obstruction of right ventricular outflow and inflow caused by the pulmonary valvular stenosis, the hypertrophy, and the presumably poor compliance of the anatomic right ventricle, however, was sufficient to cause shunting at the atrial level. Contraction of the atriialized portion of the anatomic right ventricle may also have caused shunting. In contrast, the left atrial diaphragm, although anatomically impressive, clearly was not obstructive in view of the normal pulmonary arterial pressure.

Although the point cannot be proved, it is likely that...
the patient's death resulted from paradoxical embolization of the right internal carotid artery, and that similar paradoxical emboli also caused the renal infarcts. This mode of exit has been reported in cases of Ebstein's malformation. The thrombotic material found in the right atrium in our case could well have been the source of the postulated emboli. Lack of atherosclerotic changes in the vessels involved and lack of evidence of previous thrombosis or infarction in either the two organs affected or other organs support this hypothesis. It is difficult to relate the preceding cardiac catheterization to the terminal events unless in some way it precipitated the development of thrombi in either the systemic venous system or the right atrium which could then have broken off to form emboli.

REFERENCES
3 Caddell JL, Browne MJ: Right ventricular hypertension and pulmonary stenosis in Ebstein's anomaly of the heart. Am J Cardiol 11:100-106, 1963

Pulmonary Disease Associated with the Inhalation of Cosmetic Aerosols*

Dennis R. Ehrhardt, M.D.,** Changwoo Ahn, M.D.,
F.C.C.P.,† and Thomas M. Sauvers, M.D.;†

A case of pulmonary disease due to cosmetic aerosol inhalation in a 43-year-old woman is presented. A hypersensitivity mechanism for the disease is suggested with the etiologic agent being some constituent of cosmetic aerosols, such as polyvinylpyrrolidone. Methods for substantiating pulmonary disease due to cosmetic aerosols, such as specific stains, infrared spectroscopy and skin tests, are discussed.

Two cases of pulmonary "thesaurosis," or storage disease, were reported by Bergmann et al1 in 1958, which they believed were due to hairspray inhalation. Twelve more cases were reported by the same group in 1962.2 The typical patient with thesaurosis presents with a slight cough and malaise, few or no physical findings, and bilateral pulmonary infiltrates on chest x-ray film. All give a history of extensive use of hairsprays. Cases in which lung biopsy was performed revealed diffuse interstitial fibrosis, alveolar lining hyperplasia, relative lack of inflammatory infiltrate and the presence of many histiocytes or macrophages containing PAS (periodic acid-Schiff) positive granules. One case also presented with areas of noncaseating granulomata resembling sarcoidosis. The discontinuance of hairspray resulted in clinical improvement and clearing of pulmonary infiltrates. The proposed etiology was a reaction of the lungs to inhaled hairspray in susceptible individuals, or more specifically, to a macromolecular resin constituent of hairspray called polyvinylpyrrolidone (PVP), which cannot be metabolized by the body. The PAS strain was supposedly positive for PVP in macrophages, and this was presented as the most characteristic finding in the entity.

Many reported cases followed, some supporting, others refuting the disease entity. Fifteen cases similar to Bergmann's were reported between 1959 and 1972. 1-10 This brought the total number of reported cases to 29.

Those who believed the entity did not exist supported their belief with much evidence and sound reasoning. PVP and hairspray inhalation experiments in animals by Draize et al,10 Calendra and Kay,11 Brunner et al12 Giovacchini et al13 and Lowmsa et al14 failed to produce lung pathology consistent with that reported for hairspray disease in humans. It was also argued by Brunner that PVP was a nonspecific strain and, therefore, could not be interpreted as specific for PVP in "thesaurosis." From experimental evidence he also contended that PVP was not PAS-positive in vitro.12 Schepers15 and Herrero16 made the point that pulmonary disease due to cosmetic aerosols might actually be a form of sarcoidosis. It was also argued that if the disease entity existed, there would be more cases reported. Several investigators also conducted surveys of hairdressers, all of which revealed no evidence of pulmonary disease consistent with that due to cosmetic aerosols.1,2,14-18

In recent years, the controversy has cooled and the case reports have dwindled. Yet the basic question, "Does pulmonary disease due to cosmetic aerosols really exist?" has never really been answered. For this reason, we are presenting the following case.

CASE REPORT

A 43-year-old Caucasian housewife was admitted to the Kaiser Foundation Hospital on May 17, 1971, with a chief complaint of fatigability, cough and shortness of breath for six weeks. Eight weeks prior to admission she had had a therapeutic abortion with no complications. Six weeks prior to admission, she noted the gradual onset of fatigability, cough

CHEST, VOL. 64, NO. 2, AUGUST, 1973

*From the Department of Surgery, Southern California Permanente Medical Group, Fontana, California.
**Department of Pathology, Walter Reed Hospital, Washington, D.C.
†Assistant Clinical Professor of Surgery, Loma Linda University School of Medicine.
‡Associate Clinical Professor of Surgery, Loma Linda University School of Medicine.

For editorial comment, see page 153

Downloaded From: http://journal.publications.chestnet.org/pdfaccess.ashx?url=/data/journals/chest/20941/ on 04/11/2017