been described, with the possible exception of the stillborn infants reported by Becker and Becker and by Dusek with right-sided juxtaposition. In both instances, multiple extracardiac anomalies were present.

Wenner proposed that left-sided juxtaposition is due to underdevelopment of torsion of the primitive cardiac tube. Dixon later suggested that right-sided juxtaposition is due to overdevelopment of the torsion process of the primitive tube. Thus, left juxtaposition has been assumed to be due to undertorsion, and right juxtaposition to overtorsion of the primitive cardiac tube. However, this generalization appears to be applicable only for the heart with the usual ventricular d-loop (noninverted ventricles). In the presence of ventricular l-loop (inverted ventricles), one may have to postulate the converse mechanism, namely, undertorsion of the primitive cardiac tube for right juxtaposition, and over-torsion for left juxtaposition (Fig 2). The latter could explain the rare occurrence of left juxtaposition in ventricular l-loop as reported by Lieberson.

Unlike the very high incidence of transposition of the great arteries in left juxtaposition, only 8 of 12 cases with right juxtaposition have shown this abnormality of the great vessels. Our case, and those of others, demonstrate that this abnormality of the atrial appendages may not necessarily be accompanied by complex intracardiac anomalies.

REFERENCES


Severe Hemolysis with a Fabric-Worn Cloth-Covered Aortic Valve Prosthesis*

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A patient developed severe hemolytic anemia one year after insertion of a cloth-covered aortic valve prosthesis (Starr-Edward No. 2320). The cloth over the three struts was disrupted but showed coverage with mostly organized collagen. Hemolysis stopped after replacement with a porcine heterograft. Fabric wear seems to augment the hemolysis in patients with cloth-covered artificial valves.

Hemolysis after prosthetic heart valve insertion is not rare. If clinically significant, a malfunction of the valve should be suspected and if possible corrected by operation. We describe a patient in whom disruption of the cloth over the three struts of an aortic prosthesis seemed to have augmented the hemolysis, necessitating its replacement.

CASE REPORT

A 20-year-old man was admitted to Northwestern Hospital on December 7, 1973, complaining of shortness of breath and dizziness on exertion for the past two months. The patient had undergone commissurotomy for a stenotic, bicuspid aortic valve in 1961. On January 16, 1973, a Starr-Edward cloth-covered aortic prosthesis size 9 (model 2320) was inserted because of evidence of aortic stenosis. The patient subsequently was placed on oral anticoagulants and later on iron substitution.

Physical examination on admission showed a pale, slightly icteric young man in no acute distress, with blood pressure of 140/70 mm Hg. Examination of the chest revealed a well-healed mid-sternal scar. An apical and a suprasternal thrill and a loud systolic ejection murmur were heard. The clicking of the prosthetic ball was not appreciably metallic. No diastolic murmur was audible. The remainder of the physical examination was noncontributory.

Complete blood count revealed: hemoglobin level of 8.9 gm/100 ml, hematocrit 20.3 percent, red cell count 1.99 million/cu mm with 33.7 percent reticulocytes and a white blood cell count of 4,800/cu mm, with a normal differential count. Numerous schistocytes were seen on the blood smear. His total bilirubin measured 2.3 mg/100 ml, his lactate dehydrogenase (LDH) 5500 mU/ml (N<200 mU/ml). He was aphaetoglobinemic. His otherwise normal urinalysis showed an iron content of 13 mg/24 hr (N<0.15 mg/24 hr).

On January 17, 1974, he was operated upon again. At operation, the aortic prosthetic suture line was found to be intact. The valve was replaced with a 23 mm porcine heterograft (Hancock). The patient was discharged eight days later on no medication. On February 15, 1974, he had a

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hemoglobin level of 12.4 gm/100 ml, hematocrit 36.8 percent, and a red blood cell count of 4.65 million/cu mm with 0.6 percent reticulocytes. His LDH measured 214 U/ml, his total bilirubin 0.5 mg/100 ml.

Examination of the excised valve showed the cloth to be totally disrupted on the inner struts (Fig 1). Though on gross examination the fabric appeared clean, there was a layer of mostly organized collagen on many, but not all, microscopic sections. This fibrous tissue, being in most places not as thick as the fabric-weave, was covered by a thin endothelial lining. In a few areas relatively fresh fibrin and platelets were seen on the surface and within the interstices of the network.

**DISCUSSION**

Cloth-covered valves supposedly decrease the frequency of thromboembolism seen with artificial heart valves, but the cloth covering has shown wear repeatedly, necessitating replacement of the prosthesis. These denuded struts not only lead to increased thrombus formation, but seem to augment hemolysis, the latter by creating more turbulences and by allowing the red cells to be damaged between two metallic surfaces. As our patient showed good tissue ingrowth on the fabric, similar to the description of Bull and Braunwald, lack of biologic acceptance of the valve is not to be blamed for the destruction of fabric or erythrocytes. The relatively small valve size in the aortic position of a physically active man might have contributed to the problems encountered here. It is noteworthy that no parabasilar leak was found, making it even more likely that the severe cloth wear increased the hemolysis.

Fabric destruction in this type of a totally cloth-covered valve is not rare and has led to the development of newer valves with composite tracks (Starr-Edwards Nos 2400 and 6400), eliminating contact between ball and tissue over the struts. This hopefully prevents problems like the one described here. We chose to replace his valve with a porcine heterograft. This abolished his hemolysis immediately.

ACKNOWLEDGMENT: We are grateful to Dr. N. D. Kostich for his review of the microscopic slides.

**REFERENCES**


**Pulmonary Granulomas in a Patient with Pulmonary Veno-occlusive Disease**


A patient with pulmonary veno-occlusive disease is described. Lung biopsy revealed noncaseating granulomas in conjunction with the typical vascular changes of this entity. This concurrence has not been previously described.

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