appendix including the mesentery contained noncaseating granulomata.

The differential diagnosis of appendiceal noncaseating granulomata must include, in addition to sarcoidosis, Crohn's disease, tuberculous enteritis and histoplasmosis. Since special stains and cultures for mycobacteria and fungi were unrevealing, it appears the latter two entities had been excluded. In addition the tuberculin and histoplasmin skin tests were nonreactive.

The differentiation between sarcoidosis and Crohn's disease is more difficult. Indeed, Kraft has recently discussed the marked histologic, immunologic, clinical and therapeutic similarities. However, since this patient had widespread sarcoidosis (heart, lungs, eye and skin) it would appear reasonable that the appendix and perhaps other areas of the gastrointestinal tract might be affected by the same disease. The histologic similarities of the two diseases are well-recognized but the lack of plasma cell infiltration in sarcoidosis has been used as a discriminating feature. In the present case the four-year history of recurrent abdominal pain was relieved by appendectomy. Since there was no surgical resection of ileum or large bowel, recurrence of abdominal pain and diarrhea or fistulae would be anticipated over the ensuing five year follow-up if the appendicitis were due to Crohn's disease. It must be pointed out that in the opinion of several authors sarcoidosis and Crohn's disease may coexist in the same patient; unfortunately, histologic confirmation of the diagnosis of sarcoidosis in two of the cases was not obtained.

The natural course of asymptomatic intestinal forms of sarcoidosis and the results of treatment with corticosteroids or other anti-inflammatory agents are presently unknown. It is noteworthy that in this case the patient was on 15 mg of prednisone on alternate days when the appendicitis occurred.

REFERENCES

Mirror-Image Dextrocardia with Situs Inversus and Coarctation of the Aorta*

A. Zelikovsky, M.D., B. Vidne, M.D., F.C.C.P., and M. J. Levy, M.D., F.C.C.P.

A rare case of mirror-image dextrocardia with situs inversus and coarctation of the aorta is briefly described. The coarctation was successfully corrected. No other anomalies were found in this patient, or among his siblings or family. This is the first patient reported in the literature known to us, in whom the association of both congenital anomalies is described.

Situs inversus has been known from ancient times. Aristotle cited two cases of transposed organs in animals, and Fabricius was the first to describe a case of transposed liver and spleen in a human being in 1600 A.D. In reviews of the literature before 1946, there were 1,032 collected cases. The frequency of occurrence varies greatly with different authors: according to Le Wald, 0.075 percent, according to Sherk, 0.003 percent.

Among more than 1,000 cases of mirror-image dextrocardia with situs inversus reported in the literature before 1946, only five cases were found in which additional intracardiac anomalies were present, an incidence similar to that of the general population. The conditions were: atrial septal defect, ventricular septal defect, pulmonary valvular stenosis, patent ductus arteriosus and tetralogy of Fallot.

The purpose of this paper is to report the first case of mirror-image dextrocardia with situs inversus associated with coarctation of the aorta.

CASE REPORT

The patient is an eight-year-old boy born in Israel to parents of Moroccan origin. Seven other siblings in the family are all healthy. At the age of four months, on routine medical examination, a diagnosis of dextrocardia was made. His growth and development were within normal limits. For

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FIGURE 1 (a). Standard leads show inverted P waves in lead I, positive P waves in aVR. Pericardial lead shows deep S waves in V5, V6 and smaller R waves with P negative, which demonstrates withdrawing from left ventricle.

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several months prior to his admission he complained of abdominal and leg pain during exercise. Physical examination on admission revealed a thin, well developed boy for his age. His blood pressure was 135/83 mm Hg in both arms. Breath sounds were normal. The apex beat on palpation was felt in the right fifth interspace in the midclavicular line, and on percussion cardiac dullness was noted on the right side. Heart sounds were normal, with a normal splitting of the second sound. A long blowing systolic murmur was audible in the right second interspace and along the spine in the midthoracic region. Peripheral pulses were normal and equally felt in both radial arteries but were weakly palpable in the femoral area, with posterior tibial and dorsalis pedis absent. An ECG showed the typical findings of mirror-image dextrocardia (Fig 1). X-ray films of the chest showed dextrocardia. Lung markings were normal and a gastric air bubble was noted on the right side (Fig 2). Brachial angiography showed dextrocardial right aortic arch, mirror image of the aortic arch vessels, and right descending aorta with subclavian and vertebral arteries dilated (Fig 3). Along the thoracic aorta, 2 cm beyond the origin of the right subclavian artery a narrow segment about 5 mm in length, with poststenotic dilatation of the thoracic aorta was demonstrated accompanied by dilated collateral vessels. A diagnosis of coarctation of the aorta with mirror-image dextrocardia with situs inversus was made.

At operation, through a right thoracotomy incision, a narrowed aortic segment, 5 mm in length, was found about 2 cm distal to the origin of the right subclavian artery. The ductus arteriosus was found obliterated. Resection of the coarctation was performed, followed by end-to-end anastomosis. Femoral, pedal and posterior tibial pulses were well palpable after operation, and the child was discharged following an uneventful recovery.

Figure 1 (b). Standard leads as in Figure 1a. Precordial leads on right side show q waves in V5, V6, tall R waves without S waves and positive P waves giving figure of left ventricle on same side. Typical ECG of dextrocardia.

Figure 2. Posteroanterior chest radiography illustrating dextroposition of heart.

Figure 3. Brachial arteriography demonstrating typical coarctation of aorta and dextroposition of heart.
**Discussion**

In mirror-image dextrocardia, which is the most common condition of right-sided heart, the anterior-posterior relationship of the various parts of the heart are normal, but their right-to-left orientation is reversed. The heart exists as a mirror image of normal. The venae cavae are on the left as is the systemic venous atrium. The venous ventricle is anterior and the systemic, posterior. The aortic arch is usually on the right. This condition is easily recognized because it is almost uniformly associated with some degree of abdominal situs inversus. The electrocardiographic changes are diagnostic: inverted P, QRS and T waves in lead I. Rarely, this condition may be associated with cardiac abnormalities.

According to Grand,

1. The associated cardiac malformations occur in 5 percent of cases and in these the most common anomalies are atrial septal defect, ventricular septal defect, pulmonary valvular stenosis, patent ductus arteriosus and tetralogy of Fallot.

Mirror-image dextrocardia with situs inversus tends to be associated with simple and correctable cardiac malformations, whereas the rest of the cardiac malpositions are usually associated with more complicated and often uncorrectable anomalies. Billig reported 17 patients with mirror-image dextrocardia and cardiac anomalies recently. Of those, the cardiac malformations were complicated or uncorrectable in only three patients.

The incidence of cardiac malformation in the general population is considered to be about 8 to 10 percent. The frequency of occurrence of coarctation of the aorta among patients with congenital cardiac disease is 6 percent, which amounts to a frequency of aortic coarctation among the general population of 1 in 6,000. If one takes the average frequency of mirror-image dextrocardia with situs inversus in the general population as 1:33,000, one can speculate and anticipate finding coarctation of the aorta in association with mirror-image dextrocardia and situs inversus occurring with an average frequency of 1:198,000,000 persons.

In other words, in our country with its present rate of 70,000 births per annum, one could expect to encounter this complex of the two anomalies in one person only once in about 2,828 years, which justified our designating it rare.

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**Congenital Subclavian Steal Syndrome: Anatomy, Physiology, Pathology and Surgical Correction**

Roque Pifaré, M.D., F.C.C.P. and Richard G. Rouse, M.D.

A 22-year old man with right aortic arch and congenital subclavian steal syndrome secondary to hypoplastic malformation of the left subclavian artery proximal to the ostium of the vertebral artery is presented. Surgical correction of the hypoplastic segment was performed with the use of knitted Dacron graft. The patient has subsequently been asymptomatic, and the graft is patent as demonstrated by postoperative aortography.

The term subclavian steal syndrome was coined in 1961 after Reivich and co-workers reported two such cases. Early cases of this syndrome were reported in adults with subclavian obstruction secondary to atherosclerosis. However, in 1963 Massumi presented the case of a six-year old boy with arteriographic evidence of congenital subclavian steal but without signs or symptoms of cerebral vascular insufficiency. In the litera-

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**Figure 1.** Aortic root injection via the right subclavian artery demonstrates a right aortic arch with a left common carotid, right common carotid and right subclavian arteries originating from the aorta but no visualization of the left subclavian artery.