

CLINICAL CONFERENCE

Cyanosis and Pulmonary Disease in Infancy

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In recent years there has been growing interest in pulmonary disease in infancy, especially in the newborn period. Therefore, it has become more important to try to differentiate primary pulmonary disease as a cause of cyanosis in the newborn period, from congenital heart disease as a cause of the same symptoms.

Since recognition of such clinical entities as hyaline membrane disease, plasma cell pneumonia and some types of interstitial fibrosis of the lung is occurring in younger and younger patients, one may get the impression from the literature that it is fairly easy to separate primary pulmonary disease from pulmonary disease secondary to congenital heart disease.

I would like to show you two children, one of whom was considered to have congenital heart disease and was found at postmortem examination to have primary pulmonary disease. In the other case the child was considered to have primary pulmonary disease and then was found to have pulmonary disease secondary to heart disease.

The first case is that of a girl who was first seen in this hospital as an infant because of a mild degree of cyanosis. She had been getting along quite well, but had been noted to breathe a little harder than the other children.

Aside from the cyanosis there was no abnormality on examination of this baby except that the second heart sound was very loud. There was no distinct cardiac murmur during the period of infancy. However, as the child was followed, a systolic murmur became audible. This was a soft, Grade II to Grade III pansystolic murmur and was maximum in the third left interspace medial to the apex. The second sound was narrowly split, fixed and was quite sharp. The electrocardiogram showed right ventricular hypertrophy. The roentgenogram at

the time showed that the heart was only slightly enlarged and that there was a prominence in the pulmonary artery area. This was a number of years ago, and at that time, the clinical diagnosis was Eisenmenger syndrome; that is, a ventricular septal defect with pulmonary vascular obstructive changes and high pulmonary artery pressure.

This child was followed for a number of years, and finally, at the age of 8 years, came to this hospital in extremis. She was quite cyanotic. She still had a faint systolic murmur, and a loud snapping second heart sound.

Roentgenograms showed that the heart was now quite large. The pulmonary artery was still quite prominent, and the major branches of the pulmonary artery also seemed to be somewhat enlarged. The vascular markings in the lung fields were essentially normal, and the most important clinical impression one got of this child was that she was in right heart failure and was cyanotic.

The diagnosis was the syndrome of pulmonary vascular obstruction, probably associated with a ventricular septal defect or patent ductus arteriosus.

The patient began to have "black-out" episodes. She would suddenly become extremely pale and ashen and would become unconscious for brief periods. These spells became increasingly severe, until she died during one of them.

At postmortem examination, the heart was normal except that the right ventricle was thick and dilated. Examination of the lungs revealed a marked pulmonary vascular sclerosis involving the media and intima of the large and medium size arteries. There was a small probe-patent foramen ovale to explain the site of right to left shunting. This was clearly a case of primary pulmonary vascular disease.

The second patient is a 2-month-old infant

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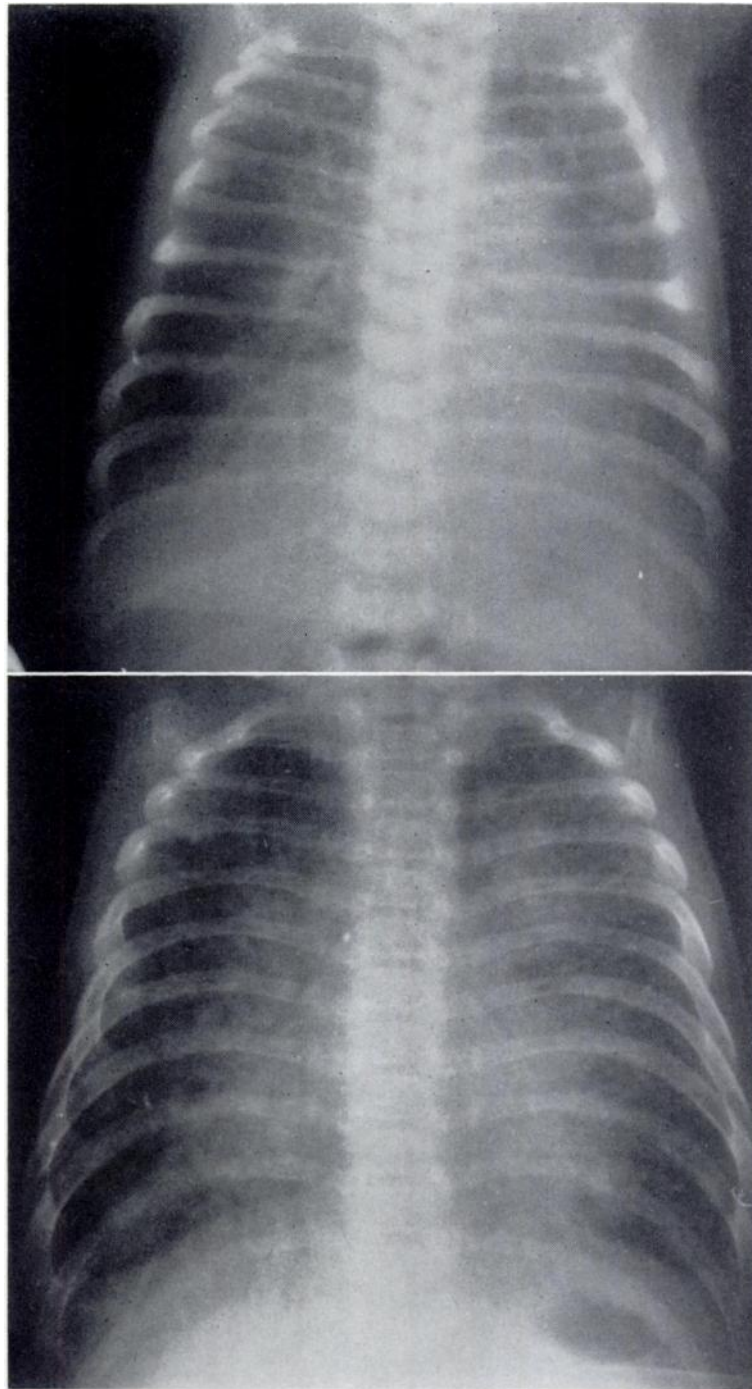


FIG. 1. (A, *upper*) Age—3 days. Note the diffuse hazy infiltrate throughout both lung fields. (B, *lower*) Age—2 months. Heart size normal. Bilateral infiltrate has become somewhat denser.

who had appeared normal at birth and was acyanotic for the first 3 days of life. The mother felt that the baby probably breathed a little harder than her other children. However, on the third day of life the baby did become mildly cyanotic. He was carefully examined, and it was found that the heart was on the right side. The people examining the baby at that time felt that with the cyanosis and dextrocardia, this child probably had congenital heart disease, and he was sent to Children's Memorial Hospital.

When the baby was admitted he was mildly cyanotic, but was extremely dyspneic. There was a very soft, ejection-type systolic murmur and the second heart sound was narrowly split. Roentgenogram of the chest showed, throughout both lung fields, a peculiar, minutely nodular, striated, rather hazy density (Fig. 1, A and B).

This density was so diffuse and widespread that it actually interfered with the differentiation of the heart shadow itself, but one could see that this was dextrocardia. The heart size was only slightly increased.

The child was put in oxygen and the cyanosis disappeared. As you know, in the newborn nursery, a fairly good test to determine whether a child's cyanosis is due to a right-to-left shunt, that is, cyanotic congenital heart disease, or to pulmonary disease, is to make the child cry while he is getting oxygen. A child with cyanotic congenital heart disease when stimulated and made to cry usually gets more cyanotic, whereas a child who may be slightly cyanotic because of pulmonary disease, when made to cry and therefore to ventilate a little better, often gets quite red, and the cyanosis rarely increases. This child responded in the way that one with pulmonary disease might.

Shortly after this an electrocardiogram was done, which showed probable right ventricular hypertrophy for this age, in addition to the mirror image dextrocardia. Incidentally, this was a situs inversus except that the distribution of the bronchi was normal, as demonstrated by a bronchogram.

With the relatively normal heart size, it was thought that the pulmonary haziness was not due to heart failure. A biopsy of the lung was obtained and it was reported as follows: There was widespread interstitial fibrosis. The fibrosis was active, in that there were many fibroblasts seen. The alveolar portion of the lungs ap-

peared to be fairly normal, and the pathology was mostly limited to the interstitial spaces which were full of this fibroblastic material. In addition, there was vascular sclerosis, that is, some hypertrophy of the media and intima of the fine arterioles.

This combination of interstitial fibrosis and pulmonary vascular sclerosis, is what was recently described in a paper on the Hamman-Rich syndrome occurring in infancy. Hamman-Rich syndrome has heretofore been considered to be an adult pulmonary disease. It usually follows multiple infections of the lung, and in addition to involving the interstitial spaces and the arterioles, the alveolar epithelium is often replaced—that is, the respiratory type of epithelium may be destroyed.

This case is presented to demonstrate that it requires a lot of information to prove that a disease like interstitial fibrosis of the lung is really primary.

Because the child had a dextrocardia, because he was cyanotic and the cyanosis seemed to be increasing, some further studies were done, among which were cardiac catheterization and a selective angiocardiogram.

During the cardiac catheterization it was possible to pass the catheter from the right ventricle into both the pulmonary artery and the aorta, showing that both great vessels communicated with the right ventricle or a single ventricle (Fig. 2, A).

In addition, a selective angiocardiogram was done from the apex of the right ventricle. The aorta and the pulmonary vessels visualized simultaneously.

Subsequent films on the angiocardiogram revealed the simultaneous visualization of the pulmonary veins and an anomalous vessel going down into the abdomen. The pulmonary veins and the liver visualized simultaneously (Fig. 2, B).

In other words, all the pulmonary veins, instead of emptying into the left atrium, enter a large anomalous vessel which courses down under the diaphragm and empties into the portal circulation. This is a total anomalous pulmonary venous return into the portal system. The relatively small heart size and the associated hazy lung markings are not infrequently seen in this anomaly, and in fact should suggest the diagnosis.

It is difficult to explain the histologic appearance of this child's lungs. Even at 3 days

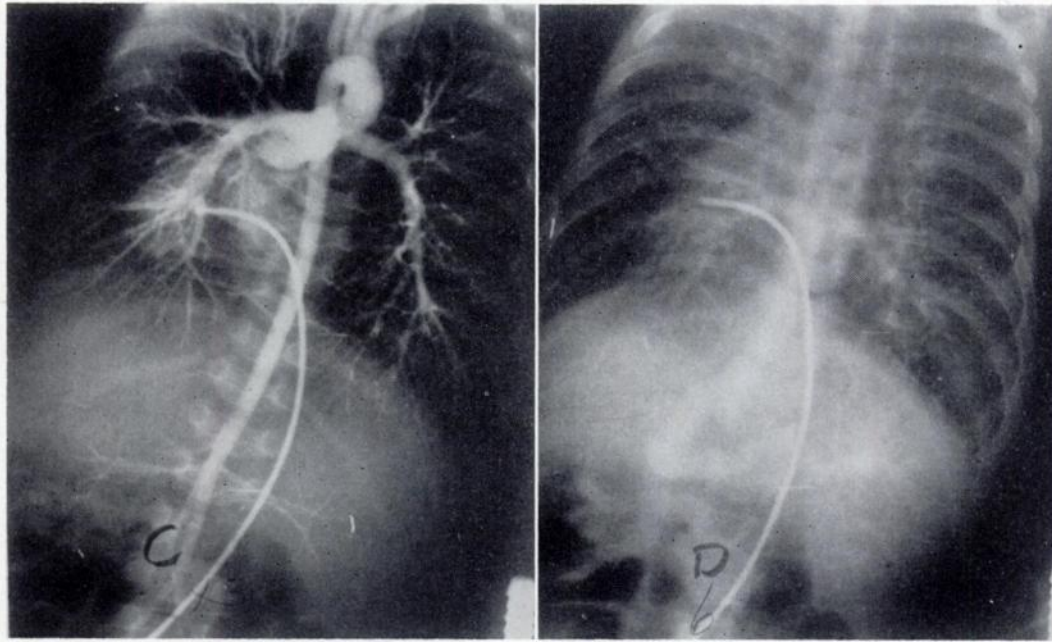


FIG. 2. (A, *left*) Selective angiogram from right ventricle. Both great vessels are visualized. (B, *right*) Two seconds after A. The pulmonary veins and the liver have been visualized. A large anomalous vessel originates in the chest, receives the pulmonary veins, descends into the abdomen and drains into the portal system.

of age, the extensive nodular infiltration was seen on roentgenogram. It is very tempting to correlate these pulmonary vascular changes and pulmonary interstitial changes with total anomalous venous drainage, possibly with some pulmonary venous obstruction during fetal life.

The child suspected of having primary pulmonary disease has to be studied completely. This baby showed the delayed onset of cyanosis and the disappearance of cyanosis in oxy-

gen that may be seen with pulmonary disease. In a sense, the biopsy "confirmed" this impression. Had the child expired at that time, the clinical diagnosis would probably have been primary pulmonary disease.

In summary, two children with extensive pulmonary pathology have been discussed. It is often difficult to differentiate primary from secondary pulmonary disease during infancy and childhood.

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