Abnormal Lung Pattern in a Newborn*

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The patient was the 3.0 kg product of a 38-week gestation of a 27-year-old gravida 1 para 0 woman receiving phenobarbital, 60 mg po, daily for seizure control. Physical examination revealed a cyanotic, lethargic newborn with a heart rate of 120 per minute, respiratory rate of 80 per minute, and a systolic blood pressure of 40 mm Hg in all extremities. There were rales over both lung fields. Breath sounds were better heard on the right. There was no heart murmur; the second heart sound was single. The liver was palpable 1 cm below the right costal margin. Peripheral pulses were diminished but equal. The electrocardiogram demonstrated right axis deviation and right ventricular predominance. Umbilical artery gases in 40 percent oxygen were a pH of 7.15 with a Po2 of 37 mm Hg and Pco2 of 65 mm Hg. Chest roentgenogram (Fig 1) was obtained and the infant was placed on a respirator.

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Diagnosis: Congenital Pulmonary Lymphangiectasia Secondary to Mitral and Aortic Valve Atresia

The chest film (Fig 1) revealed normal heart size with a diffuse small irregular shadow pattern throughout both lung fields. Despite continued ventilatory support with constant positive airway pressure, the hypoxemia was progressive and the infant expired at nine hours of age.

Postmortem examination revealed firm lungs with grossly dilated pleural lymphatics (Fig 2). The engorged pulmonary veins entered a small thick-walled left atrium. On opening the heart, atresia of the mitral and aortic valves was found. No evidence of a foramen ovale was noted.

Congenital pulmonary lymphangiectasia (CPL) is an uncommon cause of respiratory distress in the full-term newborn. The clinical manifestations of the lymphangiectasis are variable. The onset of tachypnea and cyanosis at birth carries a very poor prognosis.

Noonan et al have divided CPL into three main groups: in group 1, there is generalized lymphangiectasis. Pulmonary function is impaired, but major symptoms are due to involvement of the gastrointestinal tract. Anasarca is present in most cases. Hemihypertrophy and diffuse angiomatosis involving primarily the skeletal system have also been reported. Survival beyond infancy is seen most often in this form of CPL.

Group 2 includes those cases in which dilatation of the pulmonary lymphatics is postulated to be a result of obstruction in utero to pulmonary venous return to the heart. Cardiac malformations in this category include total anomalous pulmonary venous return (TAPVR), either above or below the diaphragm, with obstruction to flow, and atresia or stenosis of the common pulmonary vein. Another subgroup of cardiac abnormalities predisposing to CPL are those resulting from premature closure of the foramen ovale—the mitral-aortic atresia complexes. Prognosis in group 2 is also poor, and depends on the nature of the underlying cardiac malformation. At the present time, corrective surgery using deep hypothermia for infants with TAPVR is feasible and this group should be identified by means of echocardiography and cardiac catheterization. Success from surgical intervention may be adversely influenced by persistent pulmonary dysfunction.

Infants with primary pulmonary disease constitute group 3. Radiographic differentiation from neonatal focal hyperaeration (Wilson-Mikity syndrome) may be difficult, but the clinical picture is different, as is the age of onset of the roentgen findings. Congenital pneumonia, aspiration pneumonitis, and cystic adenomatoid malformation must also be considered in the full-term infant with this roentgen picture and early onset of respiratory distress. The bilateral diffuse involvement shown in lymphangiectasis is seldom seen in these conditions.

REFERENCES