Bronchial Circulation in Severe Multiple Peripheral Pulmonary Artery Stenosis

Case Report Illustrating the Origin of Continuous Murmur

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Peripheral pulmonary artery stenosis is being recognized with increasing frequency.1–3 Several observers4–6 have stressed the presence of a continuous murmur as a diagnostic feature, and continuous murmurs have been experimentally produced by graded constriction of a large branch of the pulmonary artery in dogs.5

This is the case report of an infant with severe multiple peripheral pulmonary artery stenosis in whom a loud, continuous murmur appeared to reflect flow through large, tortuous abnormally originating bronchial arteries rather than through the stenotic pulmonary arteries.

Case Report

J.S., a 6-month-old white girl was born at 41 weeks gestation following induction of labor for preeclamptic toxemia. During the second month of pregnancy the mother had a febrile exanthem thought to be rubella. Birth weight was 2,269 Gm. Cyanotic episodes were first observed at 7 days, 9 days, and 3 weeks of age, and a heart murmur was first heard on the ninth day. The infant was admitted to the hospital at 24 days of age. Positive findings at this time were a weight (2,057 Gm.) substantially below birth weight, a cataract in the right eye, and cardiovascular findings of a hyperdynamic cardiac impulse, loud second sound in the pulmonary area, and a continuous murmur maximal to the right of the sternum in the third and fourth intercostal spaces. The infant was slightly dusky but since there was no evidence of heart failure, it was decided to postpone hemodynamic studies. A skeletal survey revealed the typical radiographic features of chondrodystrophy calcificans congenita (dysplasia epiphysialis punctata).

The onset of cardiac failure at 3 months of age led to readmission. At cardiac catheterization a patent ductus arteriosus was identified by passage of the catheter from the pulmonary artery into the descending aorta. The main pulmonary artery pressure was 75/30 (55) mm. Hg and there was bidirectional shunting through the ductus arteriosus. The right and left pulmonary arteries were entered but the catheter could not be advanced into the peripheral lung fields. The systemic arterial saturation was 80.5 per cent.

At 4 months of age a large patent ductus arteriosus was ligated and following surgery the infant gained weight and the signs of cardiac failure regressed. The continuous murmur persisted, however, and at 5 months of age the recurrence of heart failure led to contrast visualization studies.

A retrograde aortogram (fig. 1) demonstrated normal anatomy of the aorta and major branches, but a large tortuous bronchial artery was seen to arise from the aortic arch and enter the right lung. Bronchial arteries arising in a normal fashion, but enlarged in caliber, were seen to enter the left lung. A right ventricular injection of contrast agent demonstrated multiple peripheral pulmonary artery stenoses.

Discussion

The diagnosis of uncomplicated bilateral pulmonary artery stenosis is suggested by the presence of a widely split but normally moving second heart sound and a long systolic ejection murmur heard equally in both parasternal regions, axillae, and interscapular areas. As in other forms of pulmonary stenosis,
right ventricular hypertrophy on the electrocardiogram usually correlates with the right ventricular pressure.\(^2\)

Commonly present continuous murmurs\(^2\)–\(^6\) have been ascribed to a continuous pressure gradient, with turbulence, across the stenotic areas of the pulmonary arteries.\(^5\)

A continuous murmur was particularly sought in each of 84 cases studied by D'Cruz et al.\(^1\) but none was found, although a wide spectrum of anatomy and severity was encountered. In three of four patients with severe bilateral pulmonary artery stenosis unassociated with other defects Delaney and Nadas\(^2\) noted that in addition to a well-transmitted systolic ejection murmur a continuous or to-and-fro murmur could be heard. Baum et al.\(^3\) observed continuous murmurs in two of 11 patients with stenosis of a main branch or branches of the pulmonary artery and in all of seven patients with stenosis of a peripheral branch or branches of the pulmonary artery.
The bronchial circulation is not commented on by these authors, and observed continuous murmurs have usually been considered to be the result of a continuous gradient across the stenosed areas of the pulmonary arteries. The demonstration of an abnormal systemic vessel to a lung forms important confirmatory evidence that this lung lacks adequate pulmonary arterial supply. Abnormal bronchial arteries are most frequently seen in tetralogy of Fallot, pulmonary atresia, absence or obliteration of a pulmonary artery, alveolar fibrosis and bronchiectasis, and bronchial carcinoma. The bronchial circulation can only be effective in gas exchange if the anastomoses with the pulmonary system are at prealveolar level.

We believe that in our patient the bronchial circulation was effective in gas exchange and that the anastomoses were at capillary or precapillary level because the contrast agent from bronchial arteries did not fill distal pulmonary arteries, but rather terminated in fine peripheral networks (fig. 1).

The continuous murmur maximal in the third and fourth right intercostal spaces exactly corresponded to a particularly tortuous segment of the abnormally arising bronchial artery supplying the right lung (fig. 1). We believe that this constitutes good evidence that the bronchial arteries are the site of the continuous murmur in this infant. Increased bronchial circulation may complicate the establishment of cardiopulmonary bypass for surgical correction and prior demonstration of these vessels may be of surgical significance.

Summary

In an infant with severe, multiple, peripheral pulmonary artery stenoses angiography clearly demonstrated enlarged tortuous bronchial arteries, especially in the right lung, where a major tortuosity corresponded closely to the point of maximal intensity of a loud continuous murmur. Increased bronchial circulation may be responsible for continuous murmurs in patients with severe multiple peripheral pulmonary artery stenosis.

References

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